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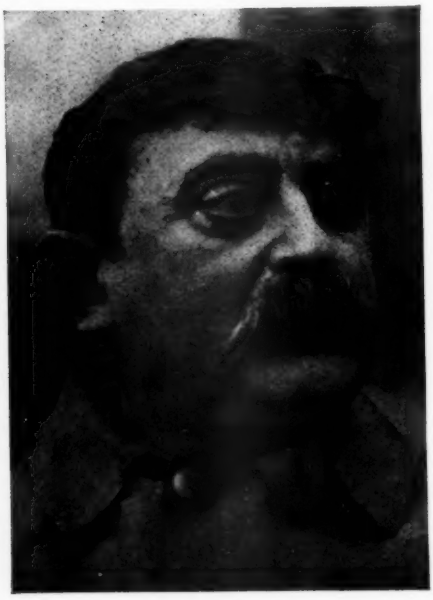
ILLUSTRATING DR. POSEY'S "REPORT OF FOUR CASES OF ORBITAL TUMORS SUCCESSFULLY REMOVED WITH PRESERVATION OF VISION THROUGH AN ORBITAL INCISION ALONG THE EXTERNAL ORBITAL RIM."



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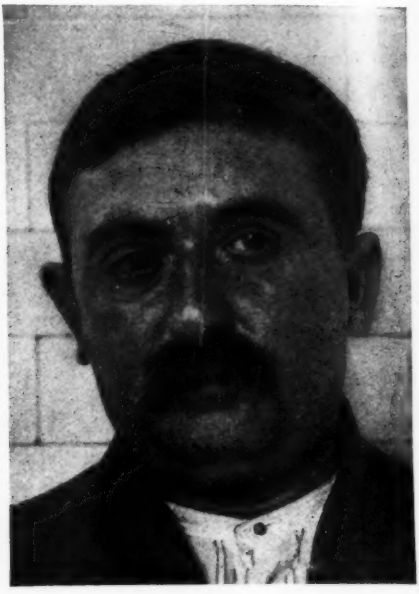

After operation.

Before operation.

After operation.

ARCHIVES OF OPHTHALMOLOGY.

REPORT OF FOUR CASES OF ORBITAL TUMORS SUCCESSFULLY REMOVED WITH PRESERVATION OF VISION THROUGH AN ORBITAL INCISION ALONG THE EXTERNAL ORBITAL RIM.[1]

BY DR. WM. CAMPBELL POSEY, PHILADELPHIA, PA.

(With four illustrations on Text-Plate XVII.)

CASE 1.—Male, 48 years of age. A continuous and gradual proptosis of the right eye past six years. No pain in globe, but occasional temporal, supraorbital, and frontal headache. No history of trauma or inflammation. Health good. Wassermann negative.

Examination.—Right eye proptosed forwards 14*mm* in advance of its fellow, with downward displacement of about 5*mm*. The globe gives the impression of being about to pop out of the orbit. No irregularity of orbital rim, no palpable mass. No pulsation. Globe cannot be pushed back into orbit. Slight bulbar injection; anterior segment of globe otherwise negative. Pupil, 3*mm*, reacts to light and on accommodation. Moderate neuroretinitis. Left eye negative. V. R. E. = $\frac{6}{6}$; L. E. = $\frac{6}{6}$. X-ray examination of orbit and surrounding sinuses negative. Nasal examination and transillumination of the sinuses negative.

Operation.—Ether. Incision over outer rim of orbit, as directed by Krönlein. After dissection of the tissues, a large encapsulated tumor, much the shape and size of the eyeball, found posterior to the eye and superior to the optic nerve, but without connection with either structure. Mass removed in

[1] Read at meeting of the American Ophthalmological Society, New London, July, 1918.

toto through the incision which was extended superiorly to give room for the exit of the tumor mass. Healing prompt, although for some days there was marked proptosis of the eyeball forwards in consequence of the hemorrhage occasioned by the trauma of the operation. Two months later, proptosis reduced to 4*mm*. Some ptosis and restriction of the outward movements of the eye. Both of these conditions are, however, rapidly subsiding. Vision as before operation. Some subsidence in the neuroretinitis.

Pathological Report.—Macroscopically the specimen consisted of a hard oval-shaped mass measuring 22 x 35*mm* in its longest diameter. The external surface was smooth. On section it was encapsulated. The tissue was white in color, resembling smooth muscle. Microscopically the tumor was encapsulated, the capsule consisting of encircling layers of long spindle cells arranged compactly in a manner resembling smooth muscle. The interior of the tumor was composed of the same type of tissue, less compactly arranged with a tendency to the formation of whirls. The blood-vessels were typical, and in most part were in the center of the whirls above mentioned. Diagnosis, spindle cell sarcoma.

CASE 2.—Male, aged 29 years. Gradual protrusion of right eye past five years. No trauma. No inflammatory symptoms.

Examination.—Right eye proptosed directly forwards 10*mm* in advance of its fellow. Health good. No traumatism. No pulsation. Impossible to press globe back into orbit. No orbital mass palpable. Globe negative. X-ray and nasal examination of orbit and accessory sinuses negative. Wassermann negative. V. R. E. = $\frac{6}{7.5}$; L. E. = $\frac{5}{5}$.

Operation.—Ether. Incision as for Krönlein procedure. Large encapsulated tumor resting upon optic nerve below found immediately behind globe without connection with adjoining structure. Readily removed by blunt dissection. Healing prompt. At end of three months, proptosis entirely gone; ocular movements free in all directions. Scar over wound almost invisible.

Pathological Examination.—Macroscopically the specimen consisted of a round mass measuring 24*mm* in diameter. It was spongy to the touch. The center consisted of a network of fibrous tissue. Microscopically the specimen was composed throughout of dense fibrous tissue, most of which was arranged in a crinkly manner, and was devoid of nuclei. The arrangement was loose, presenting many open spaces, which were lined in some instances by a very thin endothelium. The growth was spongy, the sponginess being

attributed to numerous open spaces within the aforementioned but loosely arranged fibrous tissue. Diagnosis, fibroma.

CASE 3.—Female, aged 27 years. Right eye prominent since early childhood, but no apparent increase in past ten years. Health good. Wassermann negative.

Examination.—Right eye proptosed 8*mm* in advance of its fellow. Smooth hard mass readily palpated at upper, outer angle of orbit, extending downward as far as outer commissure. V. R. E. = $\frac{6}{6}$; L. E. = $\frac{6}{6}$. Incision made over mass at outer angle of orbit. Cyst with firm capsule and dermoid contents removed without difficulty. Recovery rapid. At end of month proptosis almost entirely disappeared, some ptosis and limitation of external motion of eye.

Pathological Examination.—Dermoid cyst.

CASE 4.—Female, aged 18 years. Gradual protrusion of left eye, without inflammatory symptoms, for past eight months, this condition being attributable to an attack of typhoid fever three years previously, as a sequel of which the postcervical and submaxillary glands of the left side remained permanently swollen. One gland ruptured spontaneously. General health good. Family history negative for tuberculosis. Has never had diplopia, but covers left eye when reading to avoid seeing same line twice.

Examination.—Left eye proptosed down and somewhat out, 7*mm* in advance of its fellow. Under the upper lid at the outer third a firm movable mass, somewhat tender on firm pressure, is readily palpable, which gives the impression of being slightly lobulated and of a glandular nature. V. R. E. = $\frac{6}{6}$; L. E. = $\frac{6}{15}$. The posterior border of left sternomastoid muscle is lined with glands varying in size from a pin to a small hickory nut. Similar glands are situated in the submaxillary region. Some suggestion of enlargement of middle lobe of thyroid. Von Pirquet and Wassermann negative. Moderate anæmia. X-ray and nasal examination negative.

Operation.—Ether. Incision made along and below left supraorbital ridge at the outer part, immediately over the palpable mass. Tumor the size of a large almond and surrounded by a firm capsule exposed and removed by blunt dissection, permitting eyeball to recede into its normal position. Healing prompt. At end of a month, all proptosis had disappeared and the movements of the eye were normal.

Pathological Examination.—Macroscopically the speci-

men consisted of a hard oval-shaped mass measuring 28 x 15 *mm* in its longest diameter. It was encapsulated. On section, tissue resembled that of a gland, the center of which was hard and fibrous. Microscopically the growth was encapsulated, the capsule contributing to the formation of trabeculæ, which permeated throughout. The structure was that of an adenoma, the acini of which were small, each acinus being surrounded by a very definite connective tissue framework. The acini were about 70 microns in diameter. Near the capsule the acini arrangement was lost, probably due to compression in this region by the firm capsule. Diagnosis, adenoma.

With the exception of the fourth case, where the tumor was palpable and where the incision was naturally made directly over the mass, the precise location and the character of the neoplasm occasioning the proptosis were in doubt. The degree of the proptosis present suggested a tumor of considerable size in each instance, and the character of the displacement seemed to locate the growth in the posterior portion of the orbit. Under such circumstances it seemed advisable to enter the orbit through its outer part and to place the incision in such a way that if found desirable the outer wall of the orbit might be resected after the manner of Krönlein, thereby affording easy access to the orbital apex and permitting the easier removal of a growth from that region. Fortunately, however, in none of these cases was this found necessary. The author has performed the Krönlein procedure upon six cases, the notes of several of these having been submitted to this Society at previous meetings. All of these cases terminated successfully, and in only one was there any appreciable scarring, namely, in that of a child five years of age with a large dermoid cyst of the orbit, where the detached fragment of bone broke down, leaving for a time a discharging fistula.

All those who have performed the Krönlein operation, however, confess to the difficulty they encountered in displacing the wedge-shaped fragment of bone, resected from the outer angle of the orbit, in toto. On account of the contour of the orbital ridge and the contents of the orbit, it is impossible to employ straight saws with advantage. When chisels are employed, the bone is apt to split and it is practically impossible to remove the apex of the wedge as recommended by

the author of the operation. In several of his cases the writer has first drilled the bone with a dental engine and then employed the chisel, but even with this modification the apex of the triangle of bone has remained. Some years ago he was much impressed by an article by Magitot and Landrieu, of Paris, translated by Ziegler, of Philadelphia, which contained what appeared to be a valuable modification of the Krönlein method. As the description of this method may have escaped the attention of some of the members of this Society, it may not be amiss to quote the method in extenso. As the authors follow the original Krönlein method precisely until the resection of the bone is reached, it will only be necessary to detail their observations from that point. They are as follows:

> Now, we proceed to the bony sections. This is without doubt the most delicate stage. The reason for this is on account of the extreme hardness of the orbital border. The bony tissue of this region is certainly the most resistant of the body and its eburnation in aged subjects is normal. It is not necessary to depend upon chisels in order to finish the operation. Fortunately, we have saws. But straight saws are inconvenient. Using them horizontally, they no sooner enter the bony tissue than they injure the soft parts: for on one side are the muscles of the skin and on the other side are the eyeball and the eyelids.
>
> In order that the bony block shall reach as deep as the pterygomaxillary fissure, it is necessary that the first section shall start from this fissure, passing from the depths to the surface. Therefore, every method which commences by cutting into the orbital border only succeeds in detaching the edge of the orbit, that is to say, only a small part of the external wall.
>
> The orbital periosteum having been detached as far as the pterygomaxillary fissure, a malleable retractor is introduced into the cavity, pressing back the periosteum and the eyeball toward the nasal side. The bony border having been freely exposed by a separator (rugine), the temporal aponeurosis is incised. The object is to reach the pterygomaxillary fissure and introduce into this orifice a wire saw which should emerge through the orbit. Therefore, in order to do this, one should open up the temporal fossa a little in order to permit the passage of a probe or bodkin. With the aid of a grooved director one should then detach the whole exterior surface of the orbital apophysis of the

malar. Above all, the operator should bear in mind that it is in the angle formed by the malar apophysis and the zygoma (a great deal lower than is generally believed) that the probe must be introduced in order to penetrate into the orbit through the pterygomaxillary fissure. It is important at this point to press backward the muscular mass. It should be held back tightly by a retractor entrusted to an assistant.

Of course, one should avoid detaching the exterior periosteum of the temporo-malar, because it is through this membrane that the bony flap, dislocated and put back in place, is nourished. Then, taking a flexible bodkin, or if it is necessary, a grooved silver cannula, or better, a malleable conductor, the surgeon should give it a somewhat curved form and fasten to it the wire saw of Gigli. It is necessary then to recognize the location of the pterygomaxillary fissure. This fissure, concealed by fatty tissue, should be easy to locate with the end of a blunt instrument. It is well to note that the operator always has a tendency to look too high for it. It should be borne in mind that it is to be found at about 2.5*cm* from the orbital border formed by the *body* of the malar.

The pterygomaxillary fissure being found, the operator should introduce into it the flexible bodkin until he sees the point of the instrument emerge into the orbit, seizing it with a hemostatic forceps and drawing it toward him. He should then attach and introduce into the fissure the wire saw which he then disengages from the conductor.

There is nothing more to do now but to resect the orbital wall. The incision should be made sufficiently low in order to cut off the base of the orbital apophysis of the malar. The section, being easy, is made in a few seconds by the drill-bow movements imparted to the saw, which the two hands of the operator must hold *tightly stretched* in order to avoid kinks or breaking.

The section being finished, the surgeon searches at the extremity of the orbital apophysis of the malar for *the suture which unites the malar to the temporal*, the location being quite visible. Having recognized it he marks it with a protected saw, or in its absence, a very small straight saw, but the depth of this groove should not exceed 5 millimeters. The saw is then laid aside and the operator takes a small graver-chisel which he holds vertically, placing the cutting edge of it in the groove made by the saw. All that is necessary now is a simple blow of the mallet administered very moderately on the chisel in order to detach the *whole orbital apophysis of the malar*, as far as the pterygomaxillary fissure.

The bony block being liberated without splintering is

then *displaced* without difficulty and ultimately *put back in place* at the end of the operation.

Although the writer has practiced this method upon the cadaver, he regrets that the opportunity has not arisen to perform it upon the living subject. Although a number of cases of orbital tumor have presented themselves, the growths were removed in all with the preservation of the globe without the necessity of resecting the bone; indeed, it would appear from the author's experience that this step is necessary only in cases of tumor of the optic nerve, or in tumors situated at the apex of the orbit, which are of such magnitude that they cannot be forced between the globe and the external wall of the orbit, without injury to the former. While the modification of the Krönlein method suggested by these authors seems rational and feasible, the author confesses to having encountered great difficulty in inserting the Gigli saw through the pterygomaxillary fissure into the orbit. It is not easy to deflect the temporal muscle sufficiently to expose the fissure, and the angle at which the aneurysm needle must be made to enter the fissure is an awkward one. It is possible, however, to follow the steps outlined by the authors on the cadaver, and it may be easier, as oftentimes happens in other operative procedures, to perform it upon the living subject. The writer hopes that this method will be given a trial by others.

MICROSCOPIC FINDINGS IN A CASE OF CORALLIFORM CATARACT, WITH REMARKS ON CONGENITAL CATARACTS IN GENERAL.[1]

By Dr. F. H. VERHOEFF, Boston, Mass.

(From the Massachusetts Charitable Eye and Ear Infirmary.)

(With two illustrations on Text-Plate XVIII.)

CORALLIFORM cataract was first described as such by Marcus Gunn (1) in a brief communication to the Ophthalmological Society of the United Kingdom in 1895. His description of the clinical appearance of the cataract was as follows: "On focal illumination, rounded and oblong gray and white opacities are seen grouped towards the center of the lens, so as to resemble a piece of coral growing forwards and outwards from this point. The oblong opacities represent somewhat conical tubes, and their anterior extremities consequently appear as opaque circles or ovals. There are numerous very fine iridescent crystals throughout the clear part of the lens."

Since Gunn's report, cases of coralliform cataract have been described by Nettleship (2), Fisher (3), Langenhans (4), Kipp (5), Harman (6), and Stieren (7). Langenhans was apparently unacquainted with Gunn's case, but from his description it seems certain that his case was similar. Judging by the histories of the cases the condition is certainly present early in life, and is presumably congenital, although thus far it has not actually been observed at birth. It seems to remain almost if not quite stationary. Nettleship has shown very definitely

[1] Read at meeting of the American Ophthalmological Society, New London, July, 1918.

ILLUSTRATING DR. VERHOEFF'S ARTICLE ON "MICROSCOPIC FINDINGS IN A CASE OF CORALLIFORM CATARACT, WITH REMARKS ON CONGENITAL CATARACTS IN GENERAL."

FIG. 1.—Coralliform cataract, showing arrangement of protein crystals in lens. Hematoxylin and eosin. Photo x 11.

FIG. 2. Protein crystals in lens. Note that degenerative changes in the lens substance occur only in relation with large masses of crystals. Hematoxylin and eosin. Photo x 50.

that coralliform cataract is hereditary. He relates the case of the Betts family, to which Gunn's patient also belonged, in which thirty persons in four generations were known to have been affected. Two children of this family had lamellar cataract, whereas their fathers had the coralliform variety. Nettleship remarks that there was nothing special in the health of the Betts family. The brother of Fisher's patient had congenital cataract, and Harman reported four cases of "axial" or coralliform cataract as occurring in three generations of one family.

So far as I can ascertain, the case I have to report is the only one in which a coralliform cataract has been examined microscopically.

CASE.

Arthur B., aged 39 years, was admitted to the Massachusetts Charitable Eye and Ear Infirmary, August 9, 1912. The patient states that he first became aware at the age of ten that his sight was imperfect and that it has grown no worse since then. The visual acuity of the right eye is $\frac{5}{200}$, that of the left, $\frac{20}{100}$, and in neither eye is improved by glasses. On examination by oblique daylight illumination each lens shows the same picture. Ramnifying out from the posterior cortex is an irregular brilliantly white structure closely resembling a miniature coral. Glistening particles, supposably cholesterine crystals, are also seen here and there in the lens. The lens substance is otherwise clear and permits a fundus reflex to be seen around the opacities.

On August 13, 1912, I removed the cataract from the right eye. The usual cataract incision was made with a Graefe knife, an iridectomy done, a large piece of the anterior capsule removed with toothed capsule forceps, and the lens expressed without difficulty. Almost no cortical matter remained behind, and healing was uneventful. On August 27th, I operated on the left eye in the same manner and the patient was discharged from the hospital, September 7th. On October 4, 1912, his visual acuity in each eye with sph. $+$ 12 ⊂ cyl. $+$ 1.50 ax. 180° $= \frac{20}{100}$.

PATHOLOGICAL EXAMINATION.

Both lenses were fixed in 10% formalin. The left was retained as a macroscopic specimen, the other was embedded in celloidin and anteroposterior sections made through its

center. Each lens measured 7 by 3*mm* in size, only a small amount of the cortex evidently having been lost at operation.

Microscopic examination of the right lens reveals a remarkable picture. The lens in general is normal, but embedded in it are numerous crystals of various sizes, some of them massed together, others occurring isolated. At the posterior and anterior poles the largest masses as well as the largest crystals occur, while towards the periphery the crystals are always small in size and become more and more isolated. The crystals are confined to an area 5.5*mm* in lateral diameter, and, with rare exceptions, only in the central portion of the lens are they found at the anteroposterior limits of the specimen. They show little tendency to follow the direction of the lens fibers, often lying at right angles to the latter. Immediately around each of the larger masses of crystals the lens substance has undergone marked cataractous changes, showing vacuolization and fragmentation. Otherwise the lens substance is normal, except that here and there it shows peculiar round or oval spots, .01*mm* to .04*mm* in diameter. The central portion of each of these spots stains in hematoxylin somewhat more strongly than the lens substance while its periphery stains faintly in eosin, so that the appearance of a ring is produced. These spots occur only near the periphery of the lens and never in close approximation to the crystals or to the cataractous changes around the latter.

When favorably situated in the sections the crystals are seen to consist of rectangular or rhombic plates. The largest seen measures .44*mm* in length. The thinnest of the largest crystals as seen in the sections is .01*mm* thick, so that it is probable that this is a close approximation to their actual thickness. The smallest isolated crystals are .08*mm* long and .004*mm* thick. Generally the surface of the crystals are parallel, but occasionally one edge is twice as thick as the other. Very rarely a crystal in the shape of a hexagon is seen. This may be the result of an oblique section of a rhombic plate. The crystals injured the edge of the microtome knife only slightly, so that it was possible to obtain sections 5–8 μ thick. Numerous cross-lines, however, can be seen on the crystals, produced by the passage of the knife. Scrapings made from the cut surface of the lens also showed the crystals to consist of rhombic plates.

In sections stained in hematoxylin and eosin by the usual method, the crystals remain colorless. If, however, the sections are placed in a .2% solution of water soluble eosin in 80% alcohol for twenty-four hours they take a strong eosin stain. The crystals stain most beautifully and intensely, however, in carbol-fuchsin. By staining sections

twenty-four hours in carbol-fuchsin and then differentiating in alcohol and acid alcohol, and finally treating them with a solution of permanganate of potassium followed by oxalic acid, the crystals are stained intensely red on an almost colorless background. The crystals are also stained by Lugol's solution of iodine. They fail to stain by Gram's method. They also fail to stain in phosphotungstic acid hematoxylin. When placed in a 1% solution of osmic acid for forty-eight hours, the lens substance and the crystals are stained the same color, a light brown.

The crystals are found to be insoluble in the cold, in concentrated hydrochloric acid, sulphuric acid, acetic acid, aqua regia, ammonium hydrate, sodium hydrate, acetone, chloroform, alcohol, ether, xylol, or oil of origannm. The crystals are not melted or affected in any way when sections are boiled in water or absolute alcohol. They are destroyed when sections are placed in a saturated, aqueous solution of potassium permanganate for two hours and then in a saturated solution of oxalic acid for a few minutes. When sections are placed in concentrated sulphuric acid on a slide, and sufficient heat applied to dissolve the lens substance, the crystals are also dissolved. When a section is dried on a slide and then heated over a flame, the crystals are not destroyed until the lens substance is charred and partially burned. When sections are placed in an aqueous alkaline solution of trypsin in an incubator for more than a week, the crystals fail to dissolve. When placed in a solution of pepsin in .4% hydrochloric acid they are dissolved completely in forty-eight hours, while the lens protoplasm is only partially digested. When placed in Millon's reagent for ten minutes to twenty-four hours, the crystals are stained an intense rose color, while the protoplasm of the lens substance itself is stained much less strongly. In other words, the crystals give a strong reaction for protein by this test.

Examination of the left lens which was retained as a macroscopic specimen also reveals the presence of crystals. This lens was carried through alcohol and xylol in an unsuccessful attempt to make it more transparent. Finally the xylol was removed and the specimen placed in glycerine. When the lens is viewed under the low power of the microscope, the opacities can be seen to contain numerous transparent rhombic slates.

It is evident that the lenticular opacities in this case were due almost wholly to the presence of crystals. Compared to the crystals, degenerative changes in the lens substance were

inconspicuous, occurring almost exclusively around the large masses of crystals, and being absent about the isolated single crystals. From this it seems certain that the crystals were primarily deposited in the clear lens substance and that the degenerative changes probably resulted from their presence. The round spots described as occurring in small number at the periphery of the lens differ in appearance from any lens changes I have previously seen. They have no resemblance to the vacuoles of congenital cataracts, and never occur in close approximation to the crystals. They somewhat resemble, however, the early degenerative changes of senile cataracts, and are therefore probably of the same origin as the latter.

Owing to the arrangement of the opacities in his case, Nettleship assumed that coralliform cataract was identical in nature with the variety of congenital cataract known as spindle, axial, or fusiform cataract, and this view has been accepted by subsequent writers. In my case, similarity to spindle cataract was not noted clinically, but examination of the hardened specimens and the sections shows some indication of a spindle arrangement. Thus, the crystals are most abundant in the axis of the lens and especially at its poles, while peripherally they follow more or less closely the outline of an expanded spindle. Nevertheless, I am convinced that coralliform is entirely distinct from spindle cataract. In the first place, its coral-like appearance is so striking that the appropriateness of the term coralliform is immediately obvious. In the second place, the two microscopic examinations that have been made of spindle cataracts in rabbits by Bach and Hess have shown changes that bear no resemblance to those found in my case. Both Bach and Hess found the opacities in spindle cataract to consist of degenerated lens fibers, and no mention was made of the presence of crystals. From his examination, Hess concluded that spindle cataract was due to delayed closure of the lens sac in the embryo, whereas in my case there was no indication of any malformation of the lens.

The exact nature of the crystals is not at once obvious. Their insolubility in ether, etc., excludes the possibility of their being cholesterine crystals, although in shape they are similar to the latter. Their failure to stain in phosphotungstic acid hematoxylin excludes hemoglobin or hematoidin. Their

general insolubility and the fact that they stain in carbol-fuchsin by the tubercle bacilli method exclude all inorganic crystals. It occurred to me, therefore, that they might consist of the salt of some fatty acid, as calcium stearate. By adding calcium chloride to an alcoholic solution of stearic acid, rectangular crystals were obtained which had the same insolubility as those of the lens crystals. I was unable to stain them, however, and the possibility of the lens crystals being fatty acid in nature was of course completely excluded when I found that they gave the Millon reaction and could be digested by pepsin. That they could not be digested by trypsin is explained by their having been fixed in formalin.

Dr. E. W. Clap suggested to me that the crystals might be protein in nature and similar to protein crystals occurring in plants. It was largely due to this suggestion that the Millon test for protein was applied and found positive. Previously I had presented the problem to several physiological chemists who were unable to throw any light on the matter. Professor Osterhout suggested to me that the crystals were simply crystallized lens protein, and this suggestion seems to me certainly correct, since it explains their insolubility (after fixation in formalin), their staining reactions,[1] their positive response to the Millon test, and their digestibility in pepsin. So far as I know, this is the first instance in which protein crystals have been demonstrated in animal tissue of any kind.

The question arises as to the condition under which the crystals become deposited in coralliform cataracts. As already mentioned, Nettleship found two individuals with lamellar cataracts whose fathers had coralliform cataracts. This indicates a close relation between the two forms of cataract. Now it is well known that lamellar cataract occurs most often in children who show signs of rickets, so that it is fair to assume that this form of cataract is dependent upon a faulty calcium metabolism.[2] It seems more than probable, therefore, that

[1] I find that the protein crystals of a Brazil nut also stain differentially in carbol-fuchsin by the method for tubercle bacilli.

[2] Lamellar cataract is also sometimes associated with tetany, in which there is believed to be faulty calcium metabolism dependent upon parathyroid insufficiency. Tetany occasionally occurs in rachitic infants but it is not certain that both conditions are dependent upon the same constitutional factors.

coralliform cataract and lamellar cataract are both dependent directly or indirectly upon a faulty calcium metabolism. It is interesting to note, however, that in only one case of coralliform cataract reported, that of Langenhans, were there signs of rickets. This would tend to indicate some difference in the metabolic disturbances in the two conditions. Microscopic examinations of lamellar cataracts show that the opacities are due to the presence of fine vacuoles. It is evident that neither lamellar or coralliform cataract is dependent upon degeneration changes, as has been assumed, since in early cases such changes are absent about the vacuoles and crystals. Just what the vacuoles of lamellar cataract contain in the living lens has not hitherto been definitely determined so far as I can find, but the appearances that they present in microscopic sections indicate with considerable certainty that at some period they contained a fatty substance that has dissolved out. In this connection the following quotation from Stricker's monograph (8) seems to be of considerable significance:

> "That there is a chemical difference between the inner and the outer portions of the lens seems to be attested by a fact which, though known for a long time and frequently discussed, has been variously accounted for; namely, that in the lens of all vertebrates and young animals which during life have been perfectly transparent, immediately after death, as soon as the animal becomes cold, the inner portion of the lens becomes cloudy. Michel drew attention to the fact and showed that the cloudiness in the center of the lens of cats, pigs, and calves would disappear if they were warmed up to 15 to 20 degrees C. This could be repeated as often as desired, and each time the lens would clear up again. This cloudiness is produced by the presence of innumerable roundish, highly refracting droplets in the central portions of the lens, and is not in any way connected with the decomposition of the contents of the lens, such as is produced by freezing. In these experiments there can be no question as to the separation of the water from the albumin. These droplets become less numerous as we go from within outward. Treated with alcohol and ether, their number becomes less, and they are reduced in size. Hence it appears that they must consist of a fatty substance, which has a very low melting point, though it is impossible to state anything more definite at this time. The interest which the appearance of these bodies at a low temperature arouses is

due less to their presence than to the fact that they occur in the inner lamellæ of the lens, that their number varies in different species of animals, and that their deposit ceases a few months after birth. From this it follows that in the chemical formation of the lens, especially in the older layers of the same, very pronounced changes take place. Since, according to Kuhne and Laptschinsky, the amount of fat is greater in the old than in the young, hence it cannot be due to a quantitative but rather a qualitative change."

These observations may explain why lamellar cataract is formed only in childhood or before birth. Their possible significance in this regard, curiously enough, seems hitherto to have been entirely overlooked. In the hope of throwing further light on the matter, I recently placed the lens removed from a 3½-weeks-old human infant, which had been fixed in formalin, in 1% osmic acid for five days. The lens was first bisected to ensure penetration of the solution. On microscopic examination I found a peripheral zone, thickly studded with blackened droplets. These droplets were readily bleached when treated with a saturated solution of potassium permanganate followed by oxalic acid, thus proving that they were not ordinary fat but that they were probably related to myelin.[1] If these findings are confirmed by further observations that I hope to make, they will prove conclusively that lamellar cataract is due to the undue persistence of lipoid droplets normally present in fœtal lenses. Their persistence in cases of congenital cataract possibly could be explained by a faulty fat metabolism either dependent on or associated with the faulty calcium metabolism. Ultimately it is possible that they may be replaced by lime salts or other substances.

To explain the probable relation of coralliform cataract to faulty calcium metabolism seems difficult. Professor Osterhout informs me that the conditions under which the proteins crystallize in living tissues are not definitely known, but that almost any protein may be made to crystallize by withdrawing the salts from the solution in which it is dissolved. The possibility suggests itself, therefore, that the crystallization of the

[1] Deutschman (9), who made the first microscopic examination of a lamellar cataract, assumed, but without proof, that the droplets described by him were of myelin.

lens protein in coralliform cataract may be dependent upon a deficiency of calcium or other salts in the blood or aqueous humor.

As to the time when the crystals are deposited, it would seem that owing to the softer consistency of the fœtal lens the conditions are more suitable for their deposition in fœtal life than after birth. The great abundance and large size of the crystals seem to exclude the possibility that they are precipitated in the fluid contents of the embryonic lens cavity.

Whatever the exact natures of the droplets in lamellar cataract and of the crystals in coralliform cataract may be, it seems sufficiently clear that the lens opacities in each case result from some metabolic disturbance and are not primarily due to degenerative changes in the lens itself. We may, therefore, classify congenital cataracts into two chief types, namely, developmental cataracts, exemplified especially by posterior polar cataracts, and metabolic cataracts, exemplified by lamellar and coralliform cataracts.

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A CASE OF EVULSION OF THE OPTIC NERVE.[1]

By EUGENE M. BLAKE, Captain, M.R.C., New Haven, Conn.

IF one may judge by the paucity of references in the literature, evulsion of the optic nerve is a very rare condition. Undoubtedly the nerve is separated from the globe oftener than we realize, but because of other changes which make it impossible to examine the fundus and because the injury which produces the evulsion may be associated with grave cerebral symptoms and fatal termination, we are not permitted a great familiarity with this form of ocular traumatism. Natanson, writing in 1912, stated that his case was the sixteenth in the literature and since that time there are but two more cases, so far as I can find. The only complete description of evulsion occurs in the section of Injuries to the Optic Nerve in the *Graefe-Saemisch Handbuch*, with brief discussion in the *American Encyclopedia of Ophthalmology* and in Parsons's *Pathology of the Eye.*

The terms avulsion and evulsion appear to be used in the same sense by different authors and it would be well to employ one term consistently, rather than use two for the same condition. Avulsion is derived from the Latin *avellere*, meaning to pluck off, while evulsion comes from *vellere*, to pluck, and (*e*)—a plucking out. The latter word would seem to be the more descriptive of the two for that is what actually occurs. The optic nerve is plucked out of the globe at the point where it enters. Salzmann, in 1903, was the first to label the condition and he called it evulsio optici nervi.

The case which was seen by the writer occurred in a Russian Jew, aged 29, on May 14, 1917. Early in the morn-

[1] Read at meeting of the American Ophthalmological Society, New London, July, 1918.

ing on that date, while driving a baker's wagon, he collided with a milk team. His own wagon was overturned, throwing him to the ground in such a manner that his head struck upon a trolley track, his wagon falling on top of him. He was brought into the New Haven Hospital at 7.30 A.M. the day of the accident and the following notes recorded upon his chart.

Examination of Head and Neck.—Calvarium. Scalp wound over left parietal region near mid-line about two inches long. Closed by black silk sutures. No line of fracture or depression felt in skull. Left eye badly lacerated and bulging from orbit—enucleated in the accident room. Right eye, both lids swollen and ecchymotic. Marked conjunctival hemorrhage. Pupil slightly dilated and does not react to light, nor has patient perception of light.

Ears—negative, no bleeding.

Nose—compressed and deviated to left side. Loose from its deeper attachments. Can insert fingers into antra.

Mouth—complete fracture of maxillæ, which are loose from their attachments. Both maxillæ apparently crushed upwards and backwards. No fracture of mandible detected.

Face—wound left side chin one inch long. Wounds of mucous membrane, inner side cheeks, especially on the left side, near angle of mouth.

Chest—fracture of left clavicle about center, dislocated anteriorly. Not much over-riding.

X-Ray Report.—Fracture of both maxillæ. Possible fracture right mandible. Fracture into left orbit. Multiple fractures of maxillæ, nasal and orbital bones. Marked displacement of fragments. Possible fracture of coracoid process. Fracture left clavicle and third rib.

The physical examination upon entrance into the hospital is given in detail as it affords an explanation of the mechanism of production of the evulsion of the optic nerve in this case. The patient was thrown from his wagon, falling upon his head in such a way that the force of the blow was directed upward and to the left. This accounts for the fracture and separation of the superior maxillæ and nasal bones and the extrusion of the left eye from the orbit, as evidenced by the location of the wounds. Evidently the direct line of force was upward and to the left, but enough pressure was disseminated to the right to produce just sufficient protrusion of the right eye to separate the optic nerve from the globe without rupturing the sclera and causing collapse of the eye, as occurred on the left side. It is as though one pulled an apple from its stem.

The eye ground was examined for the first time one week after the accident. The pupil was moderately dilated and

absolutely immobile. The media were perfectly clear and the anterior chamber was of normal, or only slightly increased depth. The region of the papilla was occupied by an excavation, the depth of which was at least 8 D. but was very difficult to estimate, as there were no details to be made out at the bottom of the excavation. In color it was nearly uniformly gray, and one had the sensation of looking into a deep well.

The diameter of the hole was slightly greater than that of the normal disk. The retina was torn on the nasal side of the opening and separated from the choroid for a short distance. There was a large hemorrhage in the upper part of the retina and numerous smaller hemorrhages scattered about the fundus. A few retinal vessels appeared to have a circulation of blood through them but the greater part were white and bloodless. Veins and arteries could be differentiated with difficulty.

In the course of a few weeks the excavation at the site of the nerve head was filled in with new-formed connective tissue, the hemorrhages were absorbed and the circulation restored in several of the vessels in the lower portion of the retina. The patient made a good recovery in all respects, except of course his vision, which was nil. His temperature ranged from 97° upon admission to 101°, reaching normal on the 28th day. He was later transferred to the State Asylum for the Blind.

Evulsion of the nerve is produced in two different ways, first and the more commonly reported, by the entrance into the orbit of a foreign body such as a sharp stick or the point of an umbrella, pieces of metal, stone, or shot. Here the foreign body impinges upon the nerve, making traction backward and at the same time forcing the globe forward. The pyramidal shape of the orbit and the facility with which the globe can be pushed aside contribute to the production of this form of injury. In the second group the separation of the nerve results through the application of a crushing force, driving the eyeball forward and over-extending the optic nerve until it is pulled out of the scleral opening. The case of Reis where the upper margin of the orbit was comminuted by the kick of a horse's hoof is a good example, and the subject of the writer's case perhaps especially well exemplifies this type.

Our knowledge of the changes in these cases is slight because the number of cases which have been studied from the

beginning and for some time after is small. The separation of the optic nerve manifests itself in the early stage by a complete loss of the papilla and its vessels. At the site of the papilla is found a deep excavation. Birch-Hirschfeld found in a partial separation after a penetrating wound an excavation of 6 D. (*2mm*) and Salzmann, after a shot injury a complete separation with a *4mm* (12 D.) depression. The absence of the papilla shows as a gray or grayish blue hole. The retina may be more or less completely torn away, and if the latter, the retinal circulation is lacking. The choroid lies free, raises itself slightly, and the border of the separated retina shows a marked opacity. If the retina is not separated or only in a circumscribed place, then the retinal vessels are completely or entirely retained. The retinal circulation may be greatly increased by the proliferation of capillaries on the nervehead. Further, hemorrhages at the papillary border in the retina as well as at the macula appear. With partial separation, a deep excavation may remain behind or the excavation may be filled in by connective tissue growth, which piles up in a sheaflike deposit. Circumscribed pigment accumulation may appear.

As previously mentioned the separation of the nerve may be partial or complete. Birkhauser's patient suffered a penetration of the orbit by a cow's horn and with the ophthalmoscope he saw a small oval nervehead, with the long axis horizontal. The inferior margin was embraced by a very dark semilune which took the place of the missing portion of the nerve. There was a marked parallax between the center and the edge which left no doubt that the nerve had been partially torn loose in the lower quadrant of the scleral canal. The retinal vessels were lacerated in this direction and in six weeks the semilune had become replaced by new-formed tissue.

The dural sheath is rarely torn, as was demonstrated in the case of Dalen (*Ophthalmoscope*, vol. viii., p. 519). Microscopic sections of the eye enucleated because of glaucoma, two and one half years after the accident, showing a complete separation of the nerve but an intact dural sheath.

Experiments on cadavers were made by Hess in 1907 to produce evulsion of the optic nerve but with absolutely no success.

It would seem quite likely that in severe head injuries, such as occur in automobile accidents, train wrecks, etc., that a more careful examination of the eye ground or of the enucleated eye would show the evulsion of the optic nerve to be less rare than appears to be the case.

OCULAR AFFECTIONS DEPENDENT UPON DISEASE OF THE TONSILS: TWO CASES.[1]

By Dr. HOWARD F. HANSELL, Philadelphia.

Case 1.—Mrs. W., aged 35, was admitted to the Jefferson Hospital, May 20, 1917, under the care of Dr. F. X. Dercum. She stated that in the night of May 16th she went to bed in her usual good health. On Thursday morning, May 17th, she awoke to find the right eye entirely blind and the sight of the left eye diminished. By Saturday she was blind also in this eye. She stated that on Thursday she could see one half of objects and this half was obliterated at the expiration of four days.

At the time of my first examination she had no light perception in any part of the field of either eye. The pupils were dilated ad maximum and did not contract under strong electric light or in association with convergence. Radiographs showed no abnormality of any kind in the bones of the cranium or the contents of the skull. The sinuses of the head and face were clear. The Wassermann test of the blood and the spinal fluid was negative; when the cord was tapped the fluid was forced out through the cannula slowly and showed no sign of increased pressure. It contained no blood cells or other cells indicative of any general or special etiology. The optic nerves were swollen and a few small superficial hemorrhages were seen in the retina adjacent to the disk of the left eye. The boundary of the nerve head and the lamina cribrosa were obscured by œdema. The œdema did not extend far into the retina. The left nerve was swollen slightly more than the right. At this time there were no hemorrhages in the right eye. Examination of the urine, quantity and quality, was negative. The heart and circulatory system and the abdominal organs appeared to be normal. The patient had no nausea or vomiting; no

[1] Read at meeting of the American Ophthalmological Society, New London, July, 1918.

headache or dizziness; no pain. Indeed, the only complaint was of blindness.

She was of average height, rather fat and flabby. The only recent illness was tonsillitis, from a mild attack of which she recovered four weeks before admission into the hospital. For this affection she had been under the care of a physician.

Report of the examination of the throat by Drs. Chevalier Jackson and Fielding O. Lewis was that the tonsils were cryptic and contained broken-down tissue and probably pus, and ought to be removed. Up to this time the treatment had consisted of daily sweat baths, mercurial inunctions, and potassium iodide. No improvement whatever was apparent. Indeed, while the optic neuritis became no more pronounced, two or three hemorrhages had appeared in the retina adjacent to the nerve in the right eye. The pupils maintained their wide dilatation and light perception remained absent.

Twenty-four hours after both tonsils had been enucleated perception of light returned to the left eye, and in one day more to the right eye. The hemorrhages began to be absorbed and the swelling of the nerve head, never higher than one diopter, to subside.

During the eleven days of blindness no new symptoms developed. Mercury was badly borne and had to be discontinued. The iodide and the baths were kept up and strychnia was added. Each day after the tonsillotomy vision became better until it equaled counting fingers at six feet. The nerve heads have begun to grow pale on the temporal sides, the optic neuritis is passing on into atrophy. On June 13th daily applications of high frequency current were commenced. The return of vision has been gradual and not limited to any one part of the fields. Simultaneously the pupils began to decrease in size and the reaction to light was reëstablished.

Without following in detail the recovery of vision it may suffice to state that it was gradual, and that in March, 1918, nine months afterward, it equaled R. $\frac{5}{10}$, L. $\frac{6}{7.5}$. At this time the optic disks were pale and their vascularity was reduced. The fields of vision were not abnormally restricted for white but slightly and concentrically for colors, all of which were slowly distinguished. During the first months of recovery she was treated with strychnia, potassium iodide, high frequency current, and massage.

After eliminating every other cause by careful and thorough investigation of the other organs of the body it was concluded that the source of infection was the purulent tonsils. The rapid loss of vision in each eye and the speedy recovery after enucleation of the tonsils seemed to point unquestionably to this etiology.

The onset and cause of the blindness do not correspond to the usual history of acute axial neuritis. In this affection pain is a constant symptom in the forehead and temple or deep in the orbit, increased by movements of the eyes or by deep pressure. Vision declines rapidly. The pupils are dilated and while they do not respond to light contract consensually. With increasing loss of vision the pain grows less. The blindness is seldom complete and binocular. The fundus remains normal or there is a moderate degree of optic neuritis. After several days of treatment by sweats or inunctions the periphery of the field is restored but a large central scotoma persists.

In perineuritis or neuritis interstitialis periphera, the central vision is preserved and the field is concentrically limited for white and colors.

CASE 2.—Mrs. C., aged 30, for several weeks had repeated attacks of iridocyclitis. In the intervals the eye cleared and pain subsided. Vision remained dull. For the last five days and nights the pain has been severe and uninterrupted and she has had but a few hours' sleep. Vision equaled $\frac{3}{200}$. The cornea was stippled, anterior chamber deep, and the iris partially attached to the lens capsule. The ciliary region was excessively sensitive to touch, and all the ciliary vessels were injected. Local and constitutional remedies had been ineffective. She was admitted to the Jefferson Hospital, and double tonsillotomy for purulent disease of the tonsils was performed by Dr. Fielding O. Lewis the same day. In a few hours the pain had entirely disappeared, the synechiæ were broken off, the pupil round, and the injection decidedly less. In twenty-four hours after the operation she left the hospital practically recovered. Ten days later scarcely a vestige of the inflammation remained. This was a most striking and sudden cessation of the pathologic process, indicating beyond question that the ciliary region was being constantly fed with toxic material from the purulent tonsils.

No bacterial examination in either case was made of the pus taken from the tonsils. The infection is probably not dissimilar to that caused by abscesses around the teeth for which the staphylococci are usually held responsible.

Steinbugler (ARCH. OF OPHTHAL., March, 1918), in discussing the mode of eye infection from a dental focus, which may be identical with that of tonsillar disease, refers to Leongt's view that it takes place through channels in the bone, sub-

periosteal tissue, or by way of the lymphatics; to Polet's view that in addition to these infection may be carried through the nerves; and Dutoit adds that of the ophthalmic vein and cavernous sinuses. He asserts that a direct extension through the venous channels from the diseased area to the cavernous sinuses is anatomically possible and unquestionably has produced a sinus thrombosis and its symptoms.

E. V. L. Brown (*Ophth. Rec.*, June, 1917) reports a case of sympathetic iridocyclitis and choroiditis with preservation of useful vision in the sympathizing eye. The enucleated left eye showed typical changes. The right had thickened, tumefied iris with precipitates. Marked improvement did not occur until infected tonsils were removed, when vision improved from counting fingers to $\frac{4}{10}$, and has retained this acuteness for fourteen months.

Other cases, notably those reported by Brown and Irons (*Am. Oph. Soc.*, 1916), to the number of sixteen out of one hundred, point to the same etiology. Zentmayer (*Am. Jour. Ophthal.*, Apr., 1918) says: "That the tonsils have not more often been found to be the source of the ocular infection is surprising, when we consider how frequently they are blamed for general infection." The answer to this comment lies in the ignorance and carelessness of the oculist. In view of the great importance of focal infection in ocular disease to which the attention of the profession was called by de Schweinitz in his masterly paper read before the Ophthalmological Section of the International Medical Congress held in London in 1913, it becomes our plain duty to investigate the condition of the tonsils in every case of intraocular inflammation.

William Lang, "Etiology and Treatment of Iritis" (*Lancet*, June 23, 1917). Ten cases in private practice. In six cases there was disease of the tonsils.

I would refer those particularly interested in this subject to the paper of Dunn of Richmond, Va. (ARCH. OF OPHTHAL., Sept., 1917), who describes a number of cases of uveitis due to tonsillar disease and relieved by treatment directed to the tonsils. Such papers impress us with the necessity of diligent attention to the tonsils in inflammations of the uveal coat that are not venereal, tubercular, or traumatic.

SOME OBSERVATIONS ON THE RESTORATION OF THE ORBITAL SOCKET.[1]

By Dr. P. N. K. SCHWENK and Dr. WM. CAMPBELL POSEY, Philadelphia, Pa.

(*With seven illustrations on Text-Plates XIX. and XX. and five cuts in the text.*)

THE restoration of a socket of sufficient depth and of such configuration that an artificial eye of suitable shape may be properly fitted into it, is never an easy task, particularly when the occlusion of the socket is complete and no conjunctival lining of the lids remains. It is not the purpose of this paper to enumerate all of the methods which have been devised by operators for the accomplishment of this purpose, or to enter into comparison with the excellent results obtained by Weeks and others by the employment of Wolff and Thiersch grafts. These operations are well known and properly appreciated. The case, however, with which the lower cul-de-sac may be restored by the ingenious transposition of the skin of the cheek subjacent to the lid, into the floor of the orbit, as practiced by Maxwell, led the writers to a combination of this method with a utilization of a skin flap taken from some point above the orbit for the formation of the upper cul-de-sac. Zentmayer has successfully utilized Maxwell's procedure in forming the upper cul-de-sac, but this operation is complicated by the presence of the levator, and is not as easy of performance, nor are the results as satisfactory as when applied to the lower lid. All who have practiced plastic surgery about the orbit have experienced the rapidity with which healing occurs in this region, and the adaptability and vitality of flaps trans-

[1] Read at meeting of the American Ophthalmological Society, New London, July, 1918.

planted from parts adjoining. An exact planning of the incisions, and a precise coaptation of the margins of the flaps to the tissues with which they are brought in contact, insure prompt healing, and after a time an often undetectable scarring.

Our method of procedure is as follows: A careful separation of the margins of the two lids from one another and a free dissection of the tissues sublying. Then follows a division of the external canthus. The lower cul-de-sac is then restored by Maxwell's method and the upper by transposing a long flap taken from the skin of the forehead above the brow, care being taken, however, that no hairs of the brow are included in the margin of the flap. The edges of the flap must be nicely coaptated to the margin of the upper lid, and the upper border of the skin secured by the Maxwell procedure. The next step is of great importance, namely, the restoration of the external canthus, for unless this be accomplished at this time, the conformer which is introduced between the lids at the completion of the operation will not be retained in the cul-de-sac, and a subsequent operation for the restoration of the canthus will be necessary. To secure a firm canthus, two double needled sutures of No. 4 silk are passed through the skin of the upper pillar of the old canthus and brought out through the raw surface of the pillar; they are again reunited into the raw surface of the lower pillar, and made to issue upon the skin surface of that structure. They are then finally tied on two pearl buttons. The pedicle of the flap is thus buried under this bridge of tissue, and unless the tension upon the pillars is too great, which may be avoided by a careful dissection of the tissues of the canthal pillars prior to inserting the sutures, union is prompt and satisfactory. Should the tissues give way, the canthus may be restored later, after the flaps have united, by excising a diamond-shaped area of skin from the pedicle of the flap and uniting the edges of the denuded area in the manner just described.

Some of our colleagues have criticized our method of canthus repair, saying that a sinus would result from the burying of the skin flap under the bridge formed by the union of the two pillars. As a matter of fact, however, this has not occurred, and in the three cases about to be reported union was firm and there was no trace of a fistulous tract. Nor were we both-

ered by outgrowth of hairs from the flap. This may probably be accounted for by the youth of the patients and by the absence of hair from the tissues whence the flaps were derived. During the first few months there was considerable desquamation of skin from the flaps, but this gradually subsided and the sockets became dry and more and more commodious. A point to be accentuated is the importance of retaining the conformer in the socket for a long time, interfering with it as little as possible. The conformer being of the same form as an artificial eye, gradually molds the orbit into the shape necessary for the retention of the prothesis. A firm bandage should be kept constantly applied over the orbit for at least two weeks. The stitches inside the cul-de-sac should be removed at the end of a week, those from the canthus in eight or ten days.

CASE I.—J. A., aged 20 years, applied to the Wills Hospital for the restoration of the left socket, which had been obliterated as the result of a lime burn, as a consequence of which the burned globe had been removed some years previously. Examination showed complete anchyloblepharon, and it was impossible to insert even the smallest probe between the firmly united lids. The skin of the lids and the surrounding tissues was uninjured. The patient was etherized and after the lids had been carefully dissected apart, the external canthus divided, and the sublying tissues freely undermined, the lower cul-de-sac was restored after the method of Maxwell. A long pedicled flap was then obtained from the forehead above the brow, carefully sewed into position in the upper part of the wound, and a conformer inserted into the socket thus obtained. The canthus was then restored in the manner already described and a bandage applied.

Healing was prompt. As the patient lived at a distance, he was permitted to go home, with the conformer in excellent position. Unfortunately, however, the oculist to whom he was referred, not appreciating the importance of careful oversight over the patient, permitted him to become careless in the flushing out of the socket with boracic acid solution, in consequence of which the nasal end of the conformer slipped out of the socket and a rapid contraction of the newly found cavity resulted. Upon his return to us, it was found that the nasal sulcus of both the upper and lower cul-de-sacs had contracted, and the conformer was no longer retained under the pillars at the outer canthus. Subcutaneous incisions were made under local anæsthesia along the

lid borders and under the tissues at the canthus, and double armed sutures buried in the apices of the cul-de-sacs, being brought out upon the skin surface and tied over buttons, about 4 or 5*mm* from the ciliary borders of the lids. The conformer was again reinserted and the patient kept under close observation for a month. At the expiration of this period, a small segment of skin was excised from the somewhat redundant base of the flap, where the pedicle had been twisted in turning the flap into the orbit. An artificial eye can now be worn with comfort and the cosmetic result, apart from the immobility of the eye, is perfect.

CASE 2. R. B., male, aged 21 years, right eye and conjunctival cul-de-sac badly burned by lime in November, 1917. After enucleation, dense cicatricial bands almost entirely obliterated the socket, the inner portion of the lower cul-de-sac alone having any appreciable depth.

Ether was administered, and the lower cul-de-sac restored by Maxwell's method. Some weeks later, the external canthus was divided under ether, the upper lid freed from its cicatricial connections by dissection, and a flap which was taken from the skin of the upper lid just below the brow superimposed upon the raw surface thus obtained. The canthus was restored by the method just cited after a conformer had been inserted into the amply spacious socket. On the fifth day, however, the stitches holding the pillars of the canthus in apposition gave way, necessitating a refreshening of the edges of the flap and a reunion of the pillars some time later. Union was now permanent and a suitable sized eye is worn with comfort.

CASE 3. Male, aged 18 years, lost his right eye from a lacerating wound, demanding enucleation. In consequence of faulty technique in this operation, the upper lid was firmly bound down to the sublying tissues by dense cicatricial bands. The lower cul-de-sac was also somewhat contracted.

Ether was administered, the external canthus divided, and the lid freed of its cicatricial connections by dissection, the conjunctiva being undermined uniformly to the margin of the lower lid. The lower cul-de-sac was then formed by the method which will presently be reported, leaving a raw surface in the upper half of the socket. This was covered by a large pedicled flap taken from the skin of the upper lid just below the brow, as in the preceding case. After careful union of the edges of the flap to the surrounding tissues, and deepening of the apex of the cul-de-sac by three double mattress sutures, which were passed through the skin flap about 4*mm* from the lid margin and brought out through the skin of the brow about 6*mm* from the ciliary border, a

conformer was inserted and the canthus re-established as in the two previous cases. The raw surface marking the site of the flap was carefully covered by uniting the edges of the wound and a firm dressing applied. Recovery was uneventful, and a proper sized eye is retained without difficulty.

In the opinion of the authors the Maxwell operation is admirably adapted for the restoration of the lower cul-de-sac, as repeated trial has proved. The one objection to the procedure, however, is the tendency that the lid evinces to ectropionize at its outer half after healing has occurred. This may be partially overcome by making the flap narrower by 2 or 3*mm* than recommended by Maxwell, especially at its outer portion. The authors have not experienced the puckering in the skin below the lid, mentioned by some writers, except in one instance, where the deformity was readily overcome by excising a small wedge-shaped portion of skin, with the base up, at the point of greatest skin redundancy. As the operation seems to be but little practiced in this country, the authors have thought it might be not without interest to describe the various steps.

MAXWELL'S OPERATION.

Instruments—Scalpel, dissecting forceps, pressure forceps, glass mask, needles, and needle-holder.

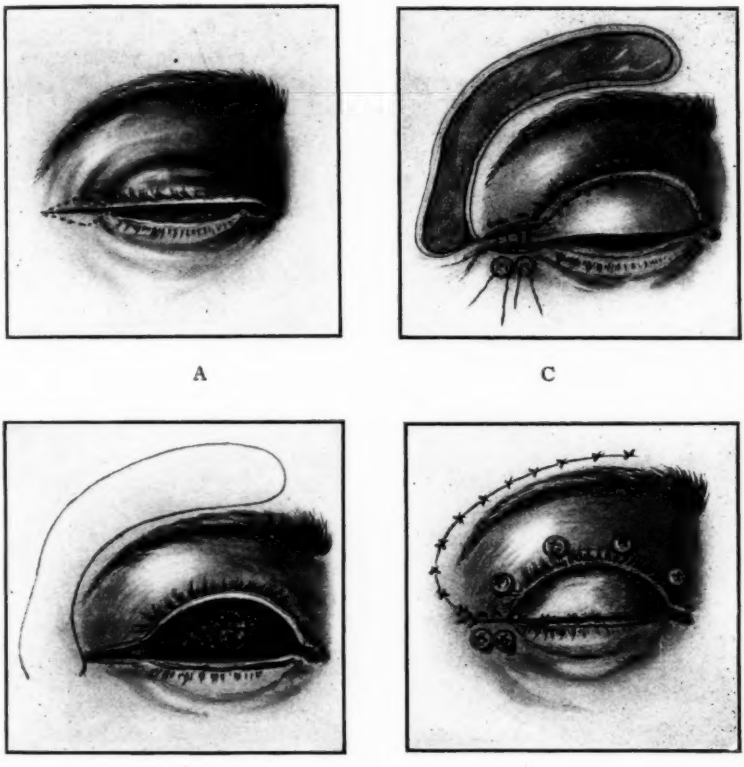
First Stage. An incision is made at the bottom of the shallow cul-de-sac in its whole length, some 5 or 6*mm* deep. It is made the same depth along all its extent (a, b, in the diagram).

Second Stage. On the skin of the lid are made two incisions, one about one sixth of an inch below the lid-margin and the other curved below this, enclosing between them a semilunar area of skin about 12*mm* broad in the widest part. The length of this curved incision (c, d, in the diagram) must be greater than that of the first. The crescent of skin thus marked out is used to form the lining of the new cul-de-sac.

Third Stage. The upper limiting incision of the crescent is deepened until it joins the incision in the cul-de-sac. The margin of the lower lid now forms a bridge attached only by its two extremities.

Fourth Stage. The crescent of skin is dissected up from its

ILLUSTRATING DRS. SCHWENK'S AND POSEY'S ARTICLE "SOME OBSERVATIONS ON THE RESTORATION OF THE ORBITAL SOCKET."



A C

B D

FIG. 1.—(A) Division of external canthus and separation of the lid margins; (B) outline of pedicled flap; (C) transposition of flap from forehead into upper portion of cul-de-sac, and restoration of external canthus; (D) coaptation of flap margins to parts adjoining.

ILLUSTRATING DRS. SCHWENK'S AND POSEY'S ARTICLE "SOME OBSERVATIONS ON THE RESTORATION OF THE ORBITAL SOCKET."

FIG. 2.—Case 1. Restoration of canthus.

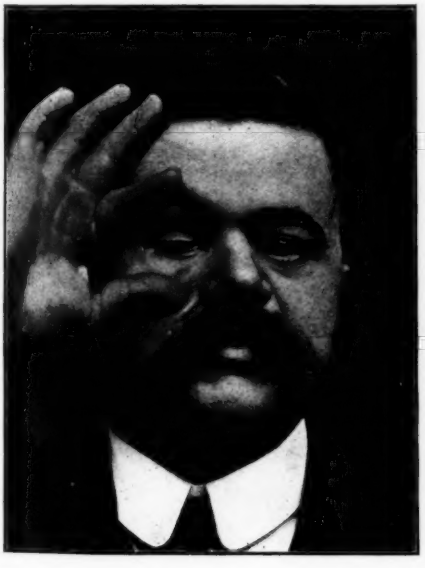


FIG. 3.—Case 4. Before operation.

FIG. 4.—Case 4. After operation.

borders until only a relatively small part (about one sixth of the whole) near the center remains attached to the subjacent tissues. This is represented as unshaded in the diagram.

Fifth Stage. The bridge of skin bearing the lashes is held forwards, and the crescent slipped under. The two extremities are secured into the angles of the first incision, and to its posterior lip the upper border of the crescent is fastened by two or three sutures. The lower border of the crescent is then brought up and stitched to the conjunctival edge of the bridge. The crescent is thus doubled on itself, and forms a groove, the posterior wall of which is made by the upper,

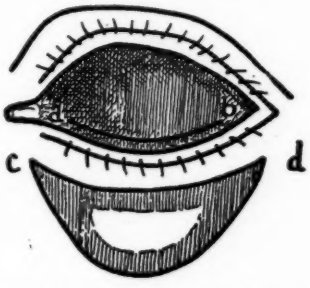


FIG. 5. Incisions for the Maxwell operation.

the anterior by the lower half of the crescent. The undissected pedicle holds the floor of the groove down.

Sixth Stage. It remains to close the wound on the surface of the lid, and this must be done carefully or there will be a troublesome scar. The two edges are very disproportionate in size: the lower much longer than the other. The first step towards closing the wound must be a stitch in the center of both edges, then one on each side midway between the center of the extremities. The coaptation of these two edges is the most difficult part of the operation.

This completes the whole procedure, but, before the pad and bandage are applied, a glass mask, preferably with a central opening, must be placed within the lids to depress the newformed cul-de-sac until healing is advanced. It should be left in for a week or ten days: the central opening is an advantage, as it enables the socket to be washed out easily.

Another ingenious method for restoration of the socket has been proposed by Golovine. In cases of exenteration of the orbit, this writer endeavored to supply a socket suitable for the reception of an artificial eye, by slipping a tongue of skin taken from the temple under a bridge of undisturbed skin at the outer aspect of the orbit. His plan of procedure was as follows: A cutaneous flap A is first shaped from the temple, as shown in the figure. A semicircular incision, a, b, is made and continued to c, through a subdermal opening, that com-

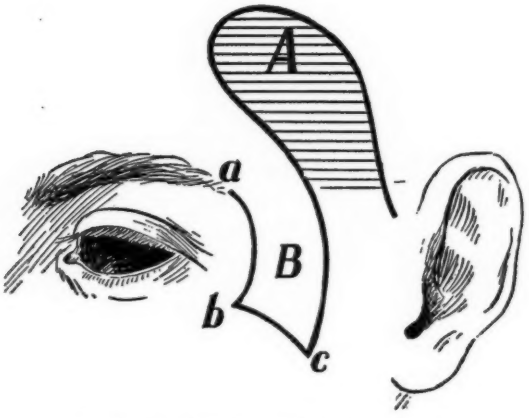


FIG. 6. Incisions for the Golovine operation.

municates with the orbit along a, b, the upper rounded edge of the large flap is slipped under, and the remaining tissues rearranged as in Fig. 7. The sliding flap applied and arranged within the orbit (epithelial surface outwards) is carefully stitched to the cut edges of the remaining conjunctiva and so a space is formed in which the prothesis may rest.

In cases where the occlusion of the socket is incomplete, some of the conjunctiva being still present, flap transplantation is not always necessary and other methods are preferable. Wiener's method is especially applicable to this class of cases, as the authors can affirm after its successful trial in several instances. As some years have elapsed since its publication,

the various steps are given in Dr. Wiener's own words, as follows:

"My method has its use where the socket, while shrunken, has still a small amount of conjunctiva left. An incision (A, B, C, Fig. 8) is made through the conjunctiva, and a flap, including only the conjunctiva, is carefully dissected down to the lid margin (A, C, Fig. 8). This dissection after being started with a knife can easily and quickly be finished with a small curved blunt scissors. The dissection is then continued with the

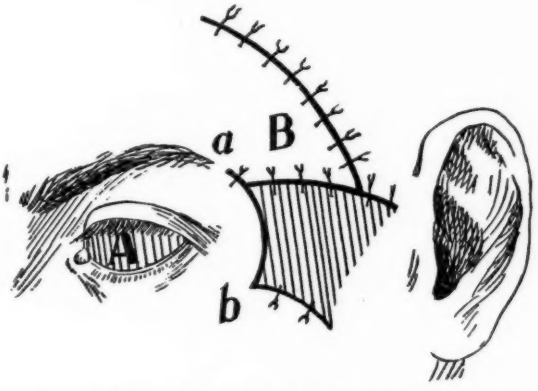
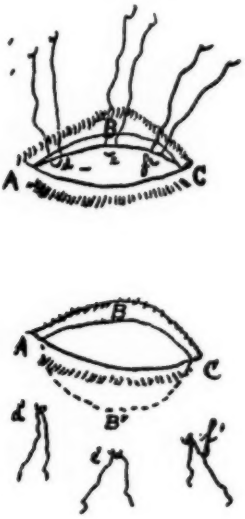


FIG. 7. Closure of the skin incisions, Golovine operation.

scissors so as to loosen the skin below the lid margin (A, C to B, Fig. 9), leaving a raw surface toward the bulbar side extended from B to B^1 and A to C (Fig. 9), and on the palpebral side A, B, C (Fig. 9). Then sutures with a needle on either end are introduced at the points d, e, f (Fig. 8), and passing them through the bottom of the newly made sulcus are brought through the skin at d′, e′, f′ (Fig. 9) and tied over a button. This gives a conjunctival covering for the lower lid and leaves the bulbar surface A, B, C, B′ (Fig. 9) to be covered. This is done by covering a lead plate, previously shaped, with grafts from the thigh and placing carefully in position. A dressing is then applied and both eyes bandaged, the patient remaining undisturbed in bed for four days, when the outside dressings are replaced by clean ones without disturbing the plate. On

about the eighth day the plate may be removed, cleaned, and replaced. The glass eye can usually be worn by the tenth or fourteenth day. The stitches at d′, e′, f′ (Fig. 9) are not removed, but are tightened each day after the fourth day until they pull through. This is an additional help to holding the lower sulcus intact, as the internal scars resulting tend constantly to pull on the bottom of the sulcus and thus heighten



FIGS. 8 and 9. Wiener operation.

the effect. It is important that these stitches should be allowed to pull through and not be removed before, for one is tempted to remove them after a week when the lower sulcus may appear too deep. I have made this mistake, thinking I had secured a lower sulcus of exaggerated depth, when, on removing the stitches, it rose too high and became almost obliterated, so that the stitches had to be replaced."

In 1915, one of the authors of this paper (Dr. Schwenk) without knowledge of Dr. Wiener's contribution, in an attempt to restore a shrunken socket, where some mucous membrane was still present, proceeded as follows:

The conjunctiva was undermined and brought forward, doubled upon itself to form a new cul-de-sac. To do this the conjunctiva was entirely loosened from the floor of the orbit from a point just below the margin of the lower lid to a point 3 or 4*mm* above the nerve head, by means of scissors, curved on the flat, entrance being made at the outer canthus. A transverse band of skin about 6*mm* wide, having its upper limits 3*mm* from the lid margin, was undermined from canthus to canthus by means of a Graefe knife inserted at the temporal side. By means of the scissors the remaining tissues of the lid were then cut through transversely into this subcutaneous pocket at a point 3*mm* from the lid margin, thus increasing the depth of the orbital cavity by an amount equal to the depth of the subcutaneous dissection. Three double-armed heavy silk mattress sutures were inserted in the loosened conjunctiva from above at a point 7*mm* from its attachment at the lid margin and the needles were carried into the subcutaneous cavity, where they were passed out through the skin at its lower part and tied on Ziegler pearl buttons; a gold conformer was then inserted.

Considerable hemorrhage resulted from cutting the hypertrophied conjunctiva, but there was no subcutaneous ecchymosis, and healing resulted without undue reaction. The stitches were allowed to remain seven days when the conformer was replaced by a glass eye.

The operation is indicated in cases in which the conjunctiva is sufficiently redundant to allow of its being slipped into the anterior cavity. It differs from the Wiener procedure in the location of the cul-de-sac anterior to the cartilage of the lid, instead of posterior to it. In the Wiener operation a large denuded area from which the conjunctiva has been taken is left exposed in the socket, which must be covered with Wolfe or Thiersch grafts. In the author's method, no raw surfaces remain, as all the dissection is done beneath skin and conjunctiva. The following case is reported as illustrative of this method.

CASE 4. M. L., male, aged 28 years. Right eye had been enucleated 11 years before on account of severe injury in a bottling establishment. Badly fitting cheap glass eyes

were worn for many years, until cicatricial contraction of socket prevented their retention. Examination showed the conjunctiva to be much thickened, corrugated, and filled with cicatrices.

Operation under ether as follows: By means of a slightly curved-on-the-flat scissors, the conjunctiva was entered at the outer canthus and two thirds of orbital mucous membrane was undermined, cutting cicatrices for some distance above the apex of the orbit and anteriorly to the margin of lower lid. Hemorrhage was pronounced, but was controlled by applications of adrenalin. A Graefe knife was then inserted just below outer canthus, and a broad band of skin was loosened and undermined, about 3*mm* from margin of the lid and 3*mm* wide parallel to its edge, across the entire width of lower lid. The submucous and the subcutaneous cavities were then united by subcutaneous incision across whole lid. Three double-armed silk mattress sutures were then inserted into the loosened conjunctiva 6*mm* from lid margin, the conjunctiva lifted from its bed on the floor of the orbit and carried with needles to the apex of subcutaneous cavity and passed through the skin externally and each tied over a pearl button to avoid puckering of the skin. The conjunctiva was carefully adapted to the lid and floor of orbit and stitched to orbital tissue to avoid sliding or displacement. The stitches thus inserted served as anchoring points and encouraged union. A gold conformer was inserted and a firm dressing was applied. The stitches were removed on the 8th day. Recovery was uneventful and the result all that could be desired, a proper sized prothesis being worn with comfort. Fig. 4 shows condition 2½ years after operation.

UNILATERAL INHERITANCE. LOSS OF THE HEREDITARY CORRELATION BETWEEN THE TWO HALVES OF THE BODY, THE PAIR ORGANS, AND ESPECIALLY THE EYES. DESCRIPTION OF TWO CASES OF THE SO-CALLED PHYSIOLOGICAL ANISOCORIA.[1]

By Dr. ANTON LUTZ, Habana.

(With six cuts in the text.)

BEAUTY is the expression of health, and is almost always based on a complete harmony of the different parts of the body. This harmony gives the impression of tranquil majesty which we receive upon contemplating ancient sculptures, such as Ludovisi's *Hera*, Otricoli's *Zeus*, Ludovisi's *Ares*, or the bust of Asclepius. We get the same impression when admiring the works of the renaissance idealistic school, of which I recall the *Mona Lisa* of Leonardo da Vinci, the *Madonna of Brügge*, of Michael Angelo, and especially the *Sistine Madonna* of Raphael. On the other hand, the Netherlands realistic school very often takes advantage of an asymmetry to obtain a humorous effect, as, for instance, Teniers in his *Dentist* or his *Alchemist*. Michael Angelo uses very seldom a complete harmony, preferring asymmetries to represent vigorous movement in his works. In the same manner, caricaturists exaggerate the asymmetry of the human body to produce a ridiculous effect. This may also be found in portraits of abnormal personalities; notable examples are: the self-portrait of the Spanish author Greco in his painting *Count Orgas's Funeral*, and the bust of the Grecian philosopher

[1] Presented at the Fourth National Medical Congress in Cuba, 1917.

Esopus, but more especially in the portrait of Saint Peter the Apostle, in the Saint Marcelin catacombs.

Besides there exists a certain antipathy for persons with marked asymmetry, for instance, of the face; they are looked upon as inferior beings, and the physician declares this asymmetry to be a sign of degeneration. A complete symmetry or harmony is very rarely found in nature. It is the origin of beauty, and it is a significant fact that divinities of all ages (even in Egypt and Babylon) have always been represented very symmetrically. This brings to our memory Raphael's words when he said that models were quite useless to him, since his works corresponded almost solely to his mental conception of beauty. Among men the rule prevails of a slight asymmetry, which may be proven by pictures of persons of all times, for example, the statues of Solon, Democrates, Sophocles, Demosthenes, of Goya's mother, etc.

Anthropologists have shown that race plays a great part, so much so that the European race shows, for example, the asymmetry of the cranium much oftener and in a more marked degree than the black race (Martin, *Anthropologie*, 1913). The reason of negroes and the insular peoples of the Pacific showing a greater symmetry of the body may be explained perhaps by the fact that they have lived during thousands of years isolated and have not had the occasion of crossing with nearby foreign elements.

Criminologists have made a special study of asymmetry of the face and of the cranium, which is to be found so often in criminals (Lombroso deems the face asymmetry as an important mark of degeneration and gives in the atlas of his work, *L'uomo delinquente*, 1888, some truly amazing instances of asymmetry of the head, also mentioning the busts of Nero and Messaline), and they have been able to verify that different sides of the face correspond sometimes exactly to those of the two parents or to those of two different individuals in the ascendency. They took a photograph of the face as seen from the front; they divided it half by half, and completed each half with its corresponding image reflected in a mirror, thus obtaining two distinct physiognomies, which resembled those of individuals in the ascendency. It seems very natural that such observations be found especially in the criminologist

literature which has time and material at its disposal. But such observations also exist in medicine. Bleuler (*Naturforschende Gesellschaft Zürich*, 1911) refers to a sage friend of his whose face corresponded on one side to that of his father, and on the other side to that of his mother.

In considering these observations the following question arises: Does a unilateral inheritance exist in such a form that one half of the body may differ in whole or in part from the other half, and due only to hereditary agencies, that is to say, transmitted by the same germinative plasm, and not occasioned by mechanical or toxic agents during the development? To solve this problem we have two ways open (not considering experiments in the vegetable and animal worlds) namely: (1st) the photographic method, for instance, of the two halves of the face; (2d) the genealogical investigation, that is, by searching for unilateral presence of an anomaly doubtless inherited during several generations past, in the same parentage.

1st. In so far as I know, there does not exist any reliable observation on the asymmetry of the entire body occasioned solely by inherited agencies,—for instance that one half of the body corresponds to that of the father and the other half to that of the mother. But we can expect to find some day such a chimera. This hope is based on the splendid experiments of the botanist, Winkler (1907) in Tübingen, which are of the utmost importance for biology, and deserve to be known to every physician. He crossed by means of grafts two different varieties of a plant (solanum nigrum and solanum lycopersicum) and thus obtained a specimen which corresponded in its branches partly to the nigrum and partly to the lycopersicum, and having leaves one half of which were nigrum and the other half lycopersicum.

The zoölogist Arnold Lang (Zürich, 1910), did not believe it impossible to obtain experimentally (influencing the blastomeres period) animals who should correspond on one side to one variety of a frog, and on the other side to another variety of the same species. Furthermore we have a series of observations on congenital asymmetry of the entire body. Cusson gathers in his thesis (Paris, 1905), twenty-six cases of hemihypertrophy of the body; among these some are found to be

without the slightest sign of disease (elephantiasis, neurofibromatosis, vasculosis, vasometric anomaly, etc.). A microscopic

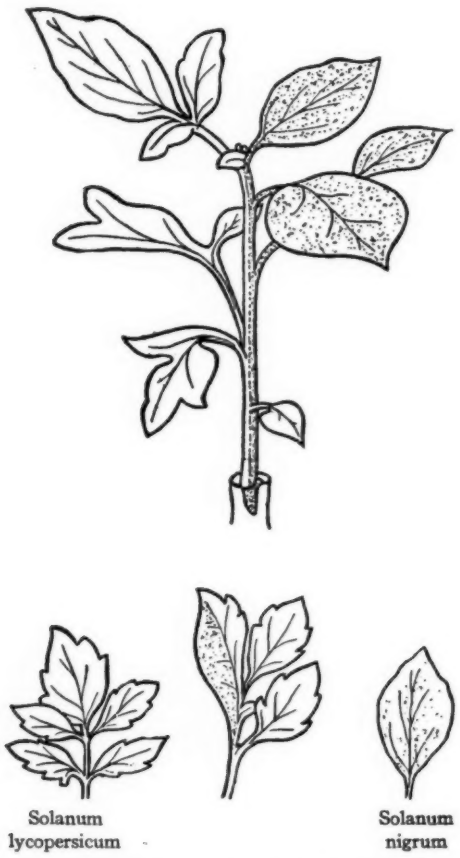


FIG. 1.—Sketches by Winkler. Taken from Arnold Lang, *Über Vererbungsversuche*, 1909.

examination showed a larger size of the ectodermal and mesodermal elements, but without the slightest pathological change, the hypertrophic half corresponding exactly to that of a normal man of larger proportions. In the description the

examination on unilateral inheritance is lacking. Of interest also are the observations of Ives, who gathered in his thesis, *Contribution à l'étude de l'œdème unilateral* (Paris, 1902), a series of œdema cases of reflex origin, which were limited to only one half of the body. The hereditary character of these œdemata has been proven many times. Two forms are known: the *acute*, of Quincke, of which Osler saw the hereditary transmission through six generations, from one to the next; and the *chronic*, or *congenital*, of Meige, of which Milroy has given us a genealogical tree of six generations (*vide* Ouvry, Thèse, Paris, 1905: *Les œdèmes familiaux*).

Magnier has in addition called attention to the fact that some hypersensitive people show on one side of the body a much greater vulnerability than on the other. He collects in his thesis (*Réactions morbides unilatérales, localisées à un seul côté du corps*, Paris, 1915) a series of observations on persons in whom the organs of one side of the body showed a tendency to sickness doubtless much greater than those of the other side.

In view of these experiments and calculations, we may hope to find some day the mulatto, who on one side of his body shall appear European and on the other African. Thus he should represent the unilateral inheritance of the characteristics of the race. One physician, a globe-trotter, who had himself a museum of medical curiosities told me that he was quite sure of having seen about thirty years ago in a medical museum at Bordeaux, a child born in due time, who showed one half of his body with the signs of the negro race and whose other half was completely white. I was not able to get more exact information about it. One observation which very nearly approaches furthermore such a probability is the following: A white girl gave birth to female twins, one of which showed all the features of the Caucasian race, whilst the other had all those of the black race. It was proven by investigations that the father of the twins was probably a negro, which conforms with the theory that both twins were originated by the same ovum, fecundated by two spermatozoa at the same time.

2d. Upon examining the different organs of the human body for unilateral hereditary anomalies, we have to consider first the skin:

A. (a) Albinism is an anomaly which is inherited in the animal and vegetable worlds in the recessive way. This leads us to hope that we may some day find Europeans, Africans, and Asiatics, who will show only one half of their body albinotic. I do not know whether there exists any reliable observation on this point. In man, observations on albinism are, in a certain degree, more difficult than in animals, because albinism is not always complete, but approaches somewhat to the light type. Furthermore, recent investigations have shown that in albinism the pigment is not only lacking in the skin, but also in the nervous system, the sympathicus, and the brain (substantia nigra). These discoveries are important for the explanation of the nystagmus, which almost always accompanies albinism of the eyes, and moreover tends to demonstrate that albinism is not only an anomaly of the skin, but of the whole ectodermal layer.

(b) Ichthyosis of the skin is observed as albinism in various persons of the same family, to wit: that it is also inherited in the recessive form (there being also cases of dominant inheritance); and we have observations on unilateral ichthyosis.

(c) Hypertrichosis is to be inherited in the dominant form, to wit, directly from the grandfather to the father, to the son, to the grandchild, etc., and normal members of the family have normal descendants. An excellent example is the Siamese family. We are still lacking observations on unilateral presence.

(d) The odor of the skin is not only characteristic of the human race, but even of all parentage. Ch. Féré calls attention (*Revue de médecine*, 1902), "l'Hérédité de l'odeur," to the point that, for instance, dogs are never mistaken in recognizing members of the same family. He cites further Hammond's observation ("The Odor of the Human Body as Developed by Certain Affections of the Nervous System," *The Medical Record*, 1877), of an hysterical woman who during her attacks perspired in only one half of her body, which unilateral perspiration emitted a violet smell.

B. Another apparatus showing hereditary anomalies is the skeleton.

(a) A hereditary anomaly whose transmission has been observed during seven generations is the polydactylia. Struthers (Edinburgh, *New Phil. Journal*, July, 1863) refers

to the following tree: "In the first generation a supernumerary finger appeared on one hand; in the second generation, in both hands; in the third, three brothers had supernumerary fingers on both hands, and one of them had also a supernumerary toe on one foot; in the fourth generation polydactylia existed in the four extremities. Unilateral polydactylia has also been observed in the cat, chicken, and horse.

(b) Drinkwater and Farabee have described a new hereditary anomaly of the skeleton which they call brachydactylia, and which consists in the absence of the first phalanx in the fingers, so that all fingers have the same structure as the thumb. Not having the original accounts at my disposal, I cannot tell whether this anomaly, which has been seen transmitted through seven (respectively five) generations, appears also on one side only.

(c) On unilateral inheritance of anomalies of the skeleton of animals. I have found some notations in Darwin (*Variation of Animals and Plants under Domestication*). He recalls Hofacker's observation, who in 1787 saw in a German forest a deer with only one horn; in 1788 two, and later on several deer were observed having but one horn on the right side. Darwin mentions also that very often sheep are to be found having on one side one horn and on the other side two horns. In the same work we find Anderson's observation on a bitch dog who had only three legs, and which bred several pups, all with the same deficiency. Anderson also recalls a genealogical tree of rabbits having but one ear.

(d) Odontology possesses as yet very few observations on hereditary transmission of anomalies. Sometimes aplasia of a tooth is inherited. Magitot (*Traité des anomalies dentaires*) saw a lady lacking from birth both lateral superior incisors, and the same defect was to be found in her daughter and in her grandchild. Quellen, M. (*Dental Cosmos*) observed the hereditary transmission of the same imperfection through three generations (grandfather, mother, and daughter). Galippe, V. (*Revue de médecine*, 1901, "Études des anomalies dentaires) observed the same imperfection, but only on one side, and also in three generations. Furthermore, he saw in a lady congenital bilateral lack of both superior incisors, whilst her daughter lacked but one.

(e) Musculature. The hereditary sickness best known of the musculature is the pseudo-hypertrophia muscularis whose hereditary character has been recognized by its first describers. Eichhorst has given us a magnificent genealogical tree of six generations (*Berliner klin. Wochenschr.*, 1873). Hoffman saw it in twin brothers, children of the same mother and same father. Nicalaysen (Virchow's *Jahresbericht*, 1876) saw it in two half-brothers, children of the same mother.

Gowers (*Lancet*, 1879) saw twin brothers of whom only one had inherited the sickness. I have been unable to find a unilateral case of this sickness. A disease still more rare of the musculature represents Thomsen's congenital myotony. Also of this we may expect to find some day a unilateral form.

C. Senses.

1. Smell. Abundo has described a hereditary form of anosmia, which is almost always mixed with hereditary hypogustation (*Rev. neurologique*, 1894); Eichhorst says that this is to be found also unilaterally (aplasia nervi olfactorii).

2. Eyes. No one has so many facilities to study the loss of correlation between the two halves of the body as the eye specialist, and consequently it is in the ophthalmological literature that we find the larger number of observations on unilateral inheritance.

(a) The most striking is perhaps the loss of the correlation of the iris color called heterochromia which had already attracted the attention of C. Darwin. In two earlier works (Lutz, Anton, *Zeitschrift f. Augenheilkunde*, 1908, and *Deutsche medizin. Wochenschr.*, 1910) I could verify Gunn's and Sym's presumption, based on a series of twenty-five cases, that the ascendance of individuals with heterochromia iridum always shows a crossing between two different colors of the iris. I could furthermore demonstrate in four families, that the child with heterochromia formed the transition between sisters with dark-colored eyes of one of the fathers, and those with the light eyes of the other of the parents. This circumstance that the child with the asymmetry is found as a transition among symmetrical children contrasts with an ancient observation based on statistics (Orchansky) to the effect that the fourth or fifth child always results better than the previous or the following ones, the loss of the correlation being doubtless an anomaly

or even a sign of degeneration. Sym (*Ophthalmic Rev.*, 1889) observed heterochromia in a child whose father had light eyes, but in one parentage all had dark eyes. In a twin child (of two ova) I observed that the father had blue eyes and the mother brown eyes, whilst one child had on one side a light brown iris and on the left a dark brown iris; the twin sister had blue irides in both eyes. Fuchs observed in twin sisters (of the same ovum) marked heterochromia: one of the twin sisters had the dark color of the father's eyes on the right side, and the light color of the mother's eyes on the left; the other twin sister showed the heterochromia iridum inversely (Fuchs, *Zeitschrift f. Augenheilkunde*, 1906). Przibram (*Archiv f. Entwicklungsmechanik*) could transmit experimentally in cats heterochromia iridum through various generations; the correlation between the light eye and deafness on the same side, which is to be found in cats, was not interrupted.

(b) Another loss of the correlation between the two eyes can be verified many times in anomalies of the refraction in anisometropia. Every eye specialist has seen cases in which one eye was emmetropic and the other one was markedly myopic or astigmatic. In four cases I was able to verify that the hypermetropia in one of the eyes corresponded to the father's eyes, and the myopia of the other eye to the mother's eyes, or inversely. The concordance of the anisometropic eyes was so very exact that they corresponded to one half of a diopter to the corresponding eyes of the parents, and one could really say that the patient had inherited in one eye the visual refraction of the father, and in the other one that of the mother. So far as I know, we have not yet a large series of observations on the relation between the anisometropic eyes of a person and those of his parents (Steiger's great work is not at my disposal), but every eye specialist will have occasion to add such new observations. These cases of marked anisometropia (for instance, hyperopia + 3.00 diopters on one eye and myopia −4.00 diopters on the other, as was shown in one of my patients) also show clearly that in the development of refraction in man, we have chiefly to blame hereditary influences, and that all other factors to which the cause was attributed have only the importance of favoring circumstances, such as working at close range of vision, an inclined position,

congestion of the cranial vessels by reason of tight collars, etc. They may be compared to the influence of climate on general pigmentation: thus, the tropical climate gives clear evidence of southern ancestors in the northern Europeans who live a long time in the tropics. If we wish to explain the development of myopia by a long pupillary distance, by the orbit's formation or by the thinness of the sclerotic, we need only state again that the condition of refraction is caused by hereditary features of the race, since specifically as to orbits we know through

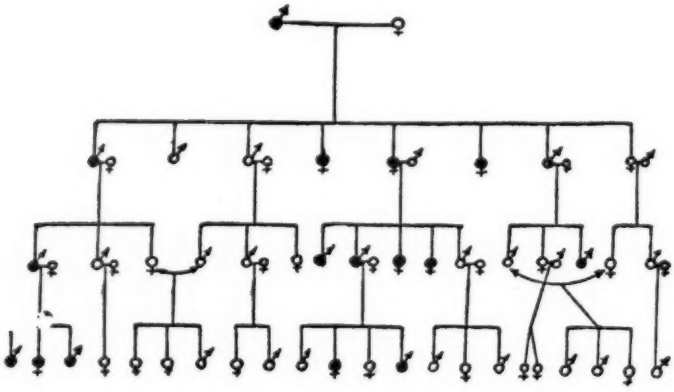


FIG. 2.—Scheme of Dominant Inheritance.

exact photographic measure that one half of the face may correspond to the mother's face whilst the other half corresponds to that of the father.

(c) Considering now the hereditary diseases of the eyes, permit me to mention briefly (more extensively, *vide* my article "Sobre la applicación del mendelismo en oftalmologia," *Anales de la Academia de Ciencias Médicas de la Habana*, January, 1912), that the modern theory of inheritance recognizes three types of hereditary transmission:

1st. Dominant inheritance: characterized in that the stigma, for instance, the disease, is inherited directly from father to son, grandchild, etc., from one generation to the next, attacking in equal number men and women, and in that consanguineous marriage has no importance. The descendants of healthy

individuals do not transmit the disease to their progeny. Figure 2 shows a scheme of the Dominant Inheritance.

2d. Sex-limited inheritance: characterized in that it is inherited only exceptionally in the direct form, from one generation to the next, but almost always skips one generation; in that men are affected ten times more than women; in that healthy men do not transmit the disease, and that sick ones transmit it very rarely; while the daughters transmit the disease as a rule without being affected themselves. Consanguineous marriage has no great importance. The following represents a scheme of sex-limited inheritance:

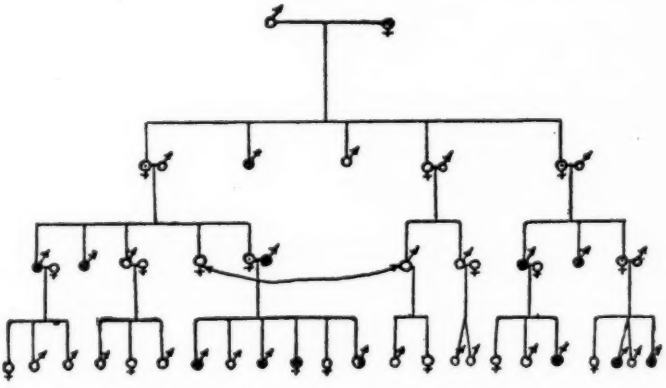


FIG. 3.—Scheme of Sex-Limited Inheritance.

3d. Recessive inheritance: characterized in that in the ascendance, sick individuals are not to be found in important numbers, but in scattered cases, sometimes many generations back; suddenly it reappears in the same family in a large number of persons, attacking indiscriminately men and women. Consanguineous marriage is of the utmost importance, this being in almost twenty-five per cent. of the cases the cause of the reappearance.

It seems difficult to give the proof of the unilateral inheritance in anomalies inherited dominantly, their prototype being cataract, glaucoma, and ptosis, because it is not always easy to mark the line between the hereditary forms and those

occasioned by other agents. But every eye specialist has seen cases in which one eye had been operated upon for cataract at a relatively early age, and the other eye remained exempt from lenticular opacity until a very advanced age. A thorough investigation of parentage will also demonstrate in this many interesting things. We have before all to eliminate the asser-

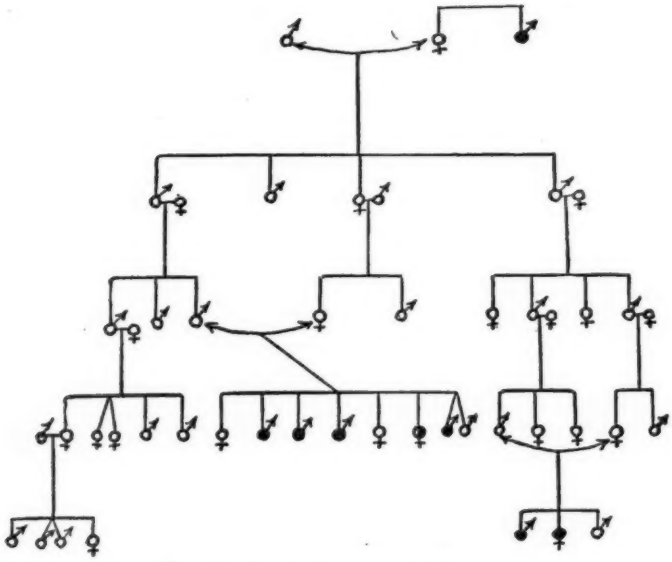


FIG. 4.—Scheme of Recessive Inheritance.

tion which is still to be found in so many manuals, that hereditary diseases of the eyes are always developed bilaterally. To-day we can only say that hereditary disease appears as a rule in both eyes (nulla regula sine exceptione).

In the sex-limited inheritance, the development of a unilateral disease has also been proven, as did Kayser with megalocornea (*Klinische Monatsbl. f. Augenheilkunde*, 1914). Another unilateral case of megalocornea combined with heterochromia was described by me in *Zeitschrift f. Augenheilkunde*, 1908.

Flesch observed unilateral megalocornea combined with angioma of the same half of the face (*Wiener mediz. Gesell-*

schaft, 1913). Von Hippel described a case of unilateral Daltonism ("Blindness as to Green and Red Colors") in Graefe's *Archiv f. Ophthalmologie*, xxvi. Other cases have been observed by Kolbe, Edridge-Green (*Lancet*, 1912), Becker (*Centralblatt. f. Augenh.*, 1888), Holmgren (2 cases in *Centralblatt. f. die med. Wissenschaften*, 1880). Hayes observed an unilateral case of Daltonism in a woman (*American Journal of Psychol.*, 1912). An asymmetry of the hereditary transmission of Daltonism has also been observed in the following forms: Reber has published that in twin boys only one was Daltonist (in both eyes). Nettleship (*Transactions Ophth. Society*, iii., 32,) saw twin sisters who were born with only one placenta and who kept their resemblance until an advanced age, and of which only one was Daltonist in both eyes. Vossius recalls, moreover, in his *Textbook of Ophthalmology* that Holmgren had also seen a patient who had on one side a normal eye, and whose other eye was blind for blue and yellow colors; but as we have not yet pedigrees on trinatopia (blindness for blue and yellow colors), such as we have on other dichromats (protanopia and deuteranopia), it is not certain whether this Holmgren case is a unilateral case, acquired or inherited. Vossius further cites Piper's observation of a patient who in one eye was blind for blue and yellow colors (tritanope), and in the other, blind for all colors (monochromate). On hereditary nystagmus which is inherited sometimes in the dominant manner and another time in the sex-limited manner, no unilateral cases are known; but I believe that it will only be a matter of time when we may find them. It will be much more difficult to give clear proof of the unilateral presence of the hereditary optic atrophy, or of the amaurosis familiaria idiotica.

In the recessive inheritance we have various valuable observations on unilateral existence. Thus, Becker has given the description of a unilateral case of congenital monochromatism ("Congenital Blindness as to All Colors," *Archiv f. Ophthalmologie*, 1879, xxv.). Vossius has described a unilateral case of retinitis punctata albescens. A large number of observations on retinitis pigmentosa unilateralis already exists in the literature of the subject. Gonin as well as Dufour (*Annales d'oculistique*, 1902–1903) and Deutschmann (*Beiträge*

z. *Augenheilkunde*, 1891) have been able to verify anatomically the unilateral existence; other observations come from Dumont, Pedraglia, Ginsberg, Anke, and Rosenbaum; Baumeister has described (Gräfe's *Archiv f. Ophthalm.*, xlx., 1873) a unilateral case of retinitis pigmentosa with deafness on the same side.

It almost seems that the larger number of cases of unilateral existence of a hereditary disease of the eyes is found in the recessive inheritance; but the number of our observations is yet too limited to tell positively in which of the inheritance forms the unilateral transmission is more frequently found.

(d) The unilateral ocular inheritance is proven not only in anatomical and physiological anomalies, but even finds its expression in such refined circumstances as the unilateral immunity. Otherwise we cannot satisfy ourselves as to these cases of unilateral trachoma, which remain during years unilateral, without any measure or precaution having been taken to avoid the infection of the second eye. Axenfeld also observed unilateral immunity in pneumococcus conjunctivitis, and even in serious blennorrhagia. During an influenza epidemic in the Maternity House, which attacked the eyes of nineteen children, I also observed a child aged twelve who had abundant secretion in only one eye for nearly ten days, and whose other eye was not infected notwithstanding that no precautions were taken.

(e) Seeing thus that the correlation of the pair organs may be lost totally or partially, unilaterally by the complicated mechanism of inheritance, we can also search for anisocoria based on unilateral inheritance. Frenkel, Henri ("Études sur l'inégalité pupillaire dans les maladies et chez les personnes saines," *Rev. de méd.*, 1879), divides anisocoria in three groups:

1st. Organic anisocoria occasioned by lesions in the different parts of the pupillary reflex way.

2d. Functional anisocoria which appears only transitorily during diseases such as pneumonia.

3d. Morphological or physiological anisocoria. Thus, it seems to me that we can only accept a physiological anisocoria, when we are able to prove that all parts of the pupillary reflex way are intact; at the present time we do not know how to make this examination with certainty. But if we should succeed in excluding all the disturbances of the pupillary reflex

path caused by diseases or congenital malformations in cases of congenital anisocoria, we should place such anisocoria equal to an anisocoria based on unilateral inheritance. Such an acceptation would be more probable if we should succeed in proving a unilateral inheritance, for instance, of the face. The physiological anisocoria is doubtless a very rare phenomenon. Reche, A. (*Deutsche mediz. Wochenschr.*, 1893) found among 14,992 cases of anisocoria, only 145 without cause, to wit: 1%. The pupillary difference between the diameter of the pupils was 0.5 to 3*mm*. Reche concludes from his abundant material that anisometropia could not be the cause of physiological anisocoria, the opinion for which Schön and Oppenheim had worked. Körbling demonstrated that the pupillary difference between an average myopic eye and an average hyperopic one was at least 0.5*mm*. Frenkel proved that an inequality of the visual acuity of both eyes could not produce a permanent anisocoria. From these considerations we deduce that the physiological anisocoria is not yet a well-defined anomaly and that care should be taken in diagnosing morphological anisocoria is seen from the following observations:

CASE 1.—P. T. José Maria, 15 years of age, laborer, very healthy, robust, well fed, without the slightest symptoms of neurasthenia or hysteria. He knows nothing about his anisocoria; he comes from a healthy family. He himself was never sick before; nor had he received a traumatism in his head. There are no lymphatic glands in the neck, paresis sympathica, pulmonary disease, dental disease (caries), nor palatal disease. Trigeminus sensibility normal. Javal in both eyes: 1°.

Sight: R. E. = refraction + 1 .50; L. E. = refraction + 1.50. Dioptric apparatus transparent. Fundi normal. Right iris slightly more clear (brown) than the left one. Anisocoria observed without the slightest change during nine months. There are no traumatic lesions of the sphincter muscle. R. E. : L. E. = 2.50*mm* : 7*mm*. Day illumination. R. E. : L. E. = 3.00*mm* : 6*mm*. Dark room. Normal reaction in both eyes: to direct and consensual light, to convergence and accommodation, to irritation of the trigeminus. Cocaine expands both pupils in the same manner, and in the same time, and first in the inferior section. After twenty minutes the following is observed: R. E. : L. E. = 7.50*mm* : 8*mm*.

The sense for colors is normal in both eyes (Nagel's test).

The visual field is normal in both eyes. The stereoscopic sense is perfect; there are simultaneous vision, stereoscopic vision, and perception of relief. There is no diplopia (examination with the red glass). There exists always a tendency to convergent squint, and then pupils show a marked myosis, the right pupil remaining nevertheless narrower.

The ocular movement is somewhat uncertain and tremulous; it is hard for the patient to distinguish objects with a firm sight. The ocular movement may be compared with ataxia in sclerosis multiplex.

CASE 2.—G. M., 34 years of age; married three years; she had two miscarriages, and has two children. She comes from a healthy family. She knows nothing about her anisocoria; she was never sick; she never received a traumatism in her head. There are no lymphatic glands in the neck, pulmonary disease, asymmetry of the face, paresis of the sympathicus, dental disease (caries), nor palatal disease. She has a tendency to obesity. Javal in both eyes: 0.50 D. Sight: R. E. = refraction + 1.00; L. E. = refraction + 1.00. Dioptric apparatus transparent. Fundi normal. Ocular movement normal in all senses; no diplopia. No enophthalmus or ptosis. Pupils: R. E.: L. E. = 6.50*mm* : 4*mm*. Day illumination. R. E.: L. E. = 7.00*mm*: 4.50*mm*. Dark room. Reaction normal in both eyes: to direct and consensual light, to accommodation and convergence, to irritation of the trigeminus. Pupils not round, but somewhat irregular as in tabes. Cocaine expands both pupils in equal manner and time. Visual field normal. Sense for colors normal. Stereoscopic sense perfect (good perception of relief).

Both of these cases present the type ordinarily called "physiological anisocoria." But upon examining them more exactly the first case shows a certain ataxy of the ocular movement, and the second one, an irregular shape of the pupil, as it often happens in tabes. This proves that the centrifugal part of the reflex pupillary way is not completely normal, and although we cannot say where the lesion is to be found, it does not seem to me fair to consider them as cases of physiological anisocoria based on unilateral inheritance. Doubtless such forms exist, since Frenkel has published some cases, but I believe that they are still much rarer than they are generally considered to be.

3. Ear. It is to Dr. Emilio Martinez, Professor of

Otology in the University of Havana, that I owe the following information on ears, taken from Heimann, Theodor (*l'Oreille et ses maladies*, 1914).

Otology recognizes three different hereditary diseases:

1st. Otosclerosis or ankylosis primaria stapediovestibularis. It is more frequently found in the feminine sex. It is inherited in the dominant manner. The inheritance has been demonstrated by Hammerschlag in four families, by Körner in three. Heimann saw a unilateral case in a physician. The physician's father, as well as two of his sons and one daughter, had the same disease; moreover a sister of the physician, two daughters, and one grandchild suffered from same.

2d. Surditis labyrinthica congenita: or congenital hypoplasia ganglii spiralis. (Alexander has demonstrated the dominant transmission and also a unilateral case.)

3d. Surdi mutitas congenita, or aplasia nerv. cochlearis. The transmission keeps an exquisitely recessive character. In at least twenty-five per cent. consanguineous marriage of the parents is to be found. It is somehow more frequently found in the masculine sex, and it is associated often with retinitis pigmentosa (Schmaltz) or albinism (Dahl). A unilateral case would represent that of Baumeister above mentioned (paragraph "recessive diseases of the eyes").

To these observations of Heimann we add the deafness of white cats with blue eyes, above cited, and whose unilateral forms have been several times observed. Darwin makes mention furthermore of an observation of Sedgwick, that persons who are color-blind from birth sometimes do not perceive musical tunes (*Medico-Chirurg. Rev.*, 1861). As unilateral Daltonism has been proven, we may suspect that there exists also a congenital impossibility to perceive the musical tunes on only one side. This syndrome of Sedgwick (Daltonism plus impossibility of perception of musical tunes) could be called "negative synopsia," in comparison with the positive synopsia (photismo: = induced sensation of color; phonismo: = induced sensation of tune), in which a certain tune awakens the impression of color, or vice versa, and whose hereditary transmission has been proven by Lomer (pedigree of four generations, *Archiv f. Psych. u. Nervenkr.*, xl.).

D. The nervous system contains a series of organic diseases,

whose hereditary character has been universally acknowledged. From these nervous hereditary diseases we must separate the neuropathic inheritance, which is almost always heteromorpha or dissimilar. Raymond says, in his *Études des pathologies nerveuses* (1910), that nervous hereditary diseases are, first, homologous, that is to say, the pathological variation always appears in the same form; they are, furthermore, homochrones, that is, appearing in the same age; and finally, homotopics, that is, the pathological change always appears in the same part of the body. Almost all of these diseases are systematized, but sometimes they affect different systems at the same time. They are abiotrophic diseases, that is to say, occasioned by atrophy of the corresponding cells of the neuron; they never are inflammatory. In the same parentage, these diseases always appear in the same manner, but the same diseases may show different variations. By organic diseases of the nervous system, whose hereditary character has been universally acknowledged, we have:

Tremor hereditarius
Chorea de Huntington
Hemicrania
Dupuytren's disease
Paramyoclonus multiplex
Ataxia cerebellaris hereditaria
Paralysis bulbaris progresiva familiaris
Ptosis familiaris tardiva (Dutil)
Infantiler Kern-schwund (Moebius)
Friedreich's disease
Paraplegia cerebralis
Menzel's disease
Atrophia spinalis muscularis hereditaria

These diseases have been acknowledged, studied, and separated, only within the last fifty years, and this explains why we do not yet have real pedigrees of them, well studied, as we have, for instance, in ophthalmology. From their complicated character it is easy to understand that the familiar anamnesis cannot help us, therefore we have left only the duty of making exact notations, that our nepotes may profit from them, as did

Keppler from those of his predecessors. As publications on these diseases are very much scattered, it has been impossible for me to find reliable observations on unilateral or asymmetrical inheritance, notwithstanding that the splendid library of Dr. José Valdés Anciano, Professor of Neurology at the University of Havana, was at my disposal, from which I have taken the majority of my notations.

Unilateral cases of ataxia cerebralis hereditaria could be: Turner's case (*Thèse de Paris*, 1856) and that of Cramer (*Beitrage z. pathol. Anatomie*, 1892).

Spiller cites (Osler's *Modern Medicine*) thirteen unilateral cases of sclerosis lateralis amyotrophica, which disease approaches very nearly the hereditary diseases of the nervous system; but these cases were all converted, later, into bilaterals.

E. By unilateral inheritance one could perhaps also explain some cases of dual personality, which is found so often in hysteria and epilepsy. This should then represent the unilateral inheritance of the psyche. Of interest also are the hallucinations. We accept to-day that they are produced by changes in the different afferent sensorial or sensitive ways, which conduce the stimuli to brains of abnormal and especially hereditary conditions, called hallucinogenous conditions. We have many observations which show that these hallucinations are sometimes exteriorized in a constant manner to only one side of the body, to one ear or to one eye (even in a hemiopic form). Wormser has in his thesis (*Sur des hallucinations unilaterales ou dedoublées*, Paris, 1895) recollected twenty-five cases out of the literature. Ziehen refers in his *Physiologische Psychologie*, 1911, a patient who suffered in one ear constantly of hallucinations of a disagreeable character, whilst he had in the other ear only others of agreeable character.

Now, as I already demonstrated in a previous article ("Über einige Stammbäume und die Anwendung der Mendel'schen Regeln auf die Ophthalmologie," von Graefe's *Archiv f. Ophthalmologie*, 1911, Bd. lxxix.) the loss of the hereditary correlation between pair organs cannot be explained in any manner by Mendelian rules which have attained such importance in the inheritance doctrine. Neither can Galton's theory give a sufficient explanation, according to which we

should have a unilateral inheritance only in cases of crossings between two ascendencies, which practically never takes place; and even in such a rare case, we would have an unilateral inheritance only in 20% (*vide* my article *Deutsche Mediz. Wochenschr.*, 1910, No. 24); such explanation would exclude almost certainly the familiar appearance of a unilateral anomaly, as, for instance, heterochromia iridum, which has been observed by Fuchs and de Schweinitz in several members of the same family. The explanation of such phenomenon can only be based on the karyokinesis.

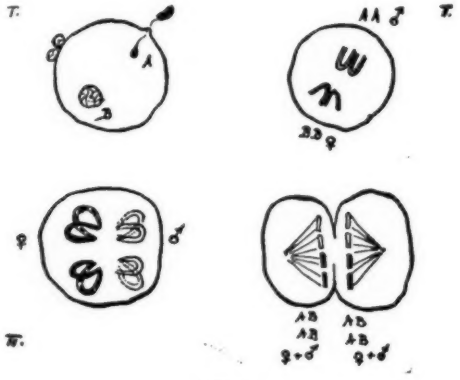


FIG. 5.

According to this theory, the bearers of the hereditary unities are the chromatin corpuscles, distributed in the chromatin loops. After the fecundation of the right ovulum by the spermatozoid, each chromatin loop is divided by the tangent in two equal ones, and the two new cells possess an equal amount of chromatin from the ovulum and from the spermatozoid. Now, by mere calculation of probability, we have to expect, that not always the same amount of AB goes on both sides (in the same manner in which a dexterous sharpshooter places his shots sometimes more to the right side of the center, and at other times more to the left). For this reason we should not be surprised to find sometimes marked asymmetries caused by the ancestral inheritance, but, on the contrary,

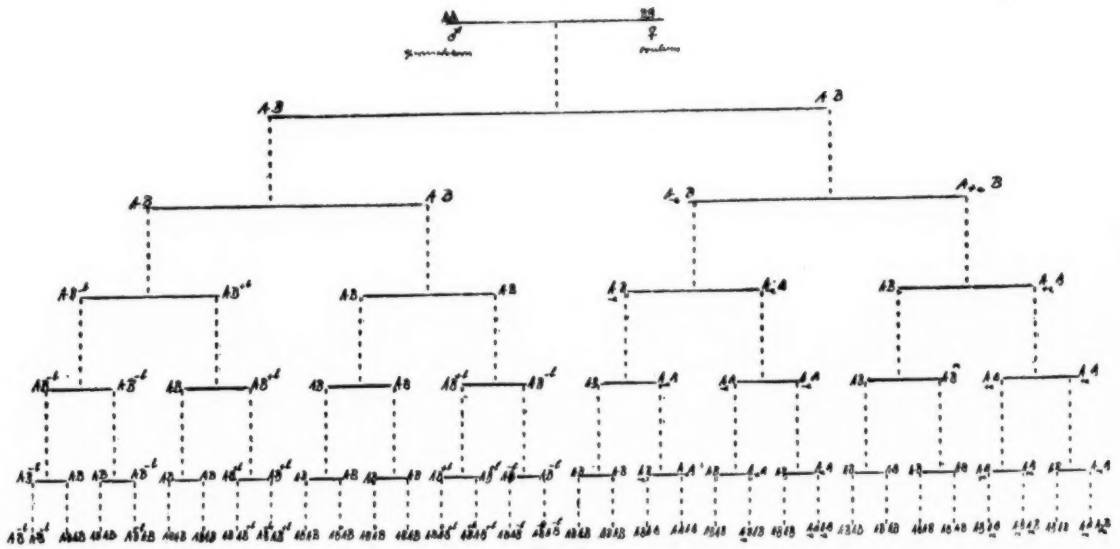


FIG. 6.

that they are not more frequently found. We have again to marvel at the precision of nature's works. These considerations explain also why we find as a rule a slight asymmetry of the face, and why we find complete harmony almost solely in deities' images. Furthermore, it explains to us why twins are so much more alike than brothers who are not twins.

The asymmetrical distribution of the chromatin corpuscles in the unilateral inheritance could be algebraically represented in the form shown in Fig. 6. This algebraical model clearly shows that the body asymmetry will be greater if the unequal distribution of the chromatin corpuscles begins early. This explains in turn why strong asymmetries, such as unilateral albinism, will be found much more rarely than, for instance, a unilateral affection of the eyes.

These demonstrations prove that by studying unilateral anomalies, we may contribute to elucidate the inheritance problems, especially in their mosaic form which Goethe so jovially described in his verses:

"Vom Vater hab ich die Statur,
des Lebens ernstes Führen,
Vom Mütterchen die Frohnatur
und Lust zu fabulieren.
Urahnfrau liebte Schmuck und Gold,
das spukt so hin und wieder;
Der Ahnherr war der Schönsten hold,
das zuckt noch durch die Glieder.
Sind nun die Elemente nicht
aus dem Komplex zu trennen,
Was ist denn an dem ganzen Wicht
noch Original zu nennen?"

ACUTE BILATERAL RETROBULBAR NEURITIS OF SEPTIC ORIGIN, RESULTING IN RAPID AND COMPLETE LOSS OF VISION.[1]

By Dr. WALTER EYRE LAMBERT, New York.

ALTHOUGH the eye history of this case, previous to the attack of retrobulbar neuritis, has no special bearing on the subject, it is in itself interesting, and I shall report it briefly.

The patient, Mrs. S., first consulted me in 1896—her age then was 44 years. She is a lady of means, and had of course most comfortable surroundings, but never seemed very well, and had a great many things to worry her. At that time she was complaining of severe headaches, which she referred to her eyes. She had always had trouble with her eyes, and was wearing glasses at the time. The refraction for the right eye was plus 1.25, and for the left eye, minus 1.25. With these glasses she had $\frac{20}{10}$ vision in each eye. I found a hyperphoria of 3 degrees. At that time I questioned whether this small amount of hyperphoria was the cause of her eye symptoms. My experience since then has shown me that small degrees of hyperphoria frequently produce asthenopia; and correcting it usually gives relief. I prescribed for her glasses with a prism of 2 degrees; these apparently gave her immediate and perfect relief, which continued for several months; when suddenly a pair of new glasses brought on a return of the headaches and feeling of eye strain. She submitted these glasses to me at once; and I found that the optician had placed the prism base up instead of down. The correction of this mistake relieved the patient immediately, and she continued to have comfortable use of her eyes for many years—with some modification of her glasses for increasing presbyopia.

[1] Read at meeting of American Ophthalmological Society, New London, July, 1918.

On March 27, 1917, the patient consulted me. I had not seen her for over a year, during which time she had been in rather poor health, suffering with severe pains in her head, and neuritis in her neck and arms. She had been under the care of several physicians in Boston; and for some time had been treated by an osteopath there, whom she felt relieved her very much. The eye condition at that time had changed but very little—a slight increase in the amount of hyperphoria, it being 4 degrees. I changed her glasses and advised her to consult her family physician in New York, who had taken care of her off and on for many years. I even then suspected the probability of some sinus trouble. An X-ray was made, the report on which is as follows: "A root fragment had been pushed into the right antrum. The antrum also shows a distinct occlusion of its air space, which possibly may have been caused by the displaced root fragment. The rest of the sinuses appeared quite normal. There is considerable constriction in the nares, caused by the deviated septum and enlarged turbinates. The sella is smaller than the average. There is no evidence of any infection about the teeth." The antrum was operated on April 6, 1917, and found to contain a considerable amount of green pus. Permanent drainage was established through the nasal cavity. The final bacteriological report of cultures from the antrum showed streptococcus viridans. The blood culture proved negative. The patient seemed to be relieved of her headaches, subsequently to this operation; although a slight septic temperature continued. She made an uninterrupted recovery as far as the operation was concerned.

On May 7, 1917, she complained of blurring of vision in the left eye. About this time the headaches had returned with some intensity, and the temperature was a little higher, which was running between 100 and 101. On May 8th, the next morning, I saw her, and found the pupil of the left eye moderately dilated, no reaction to light, and not even light perception. An ophthalmoscopic examination showed a marked, though moderate papillitis—the arteries small, and sluggish, segmented circulation in the veins. The right eye was unaffected, vision being normal. Dr. Weeks saw the patient on the same day, and agreed with me that there was partial occlusion of the central artery of the retina; probably thrombosis of septic origin.

On May 10th, the right eye became affected; a similar ophthalmoscopic picture presented itself in that eye—vision reduced to fingers. Dr. Weeks again saw the patient with me, and we agreed that an immediate operation of the ethmoid cells—which we regarded as the origin of the trouble—should be done. This was in the evening of May 10th;

the operation, however, was deferred, and the next day an X-ray was made, which gave no evidence of sinus involvement.

On May 12th the patient was seen by Dr. Knapp with me: the vision of the right eye had slightly improved, and we were able to demonstrate a distinct central scotoma for form and color, tenderness on pressure on the globe, and also on movement of the eyeball. We regarded her condition as acute retrobulbar neuritis; and the question of an operation on the ethmoid was again considered. But it was determined after very careful consideration, and consultation with two well-known rhinologists, futile. The vision of the right eye failed rapidly, and on May 15th the patient was perfectly blind. Atrophy of both optic nerves followed.

When the case was first seen, both Dr. Weeks and myself were of the opinion that there was a thrombosis of the central artery of the retina—probably of septic origin. After the right eye became involved, and we were able to demonstrate the presence of central scotoma for color and tenderness in the orbit, the diagnosis of an acute retrobulbar neuritis was made —in which diagnosis both Dr. Weeks and Dr. Knapp agreed. Nothwithstanding the negative findings of the X-ray examination, and that the ocular complications did not manifest themselves for a month after the operation on the antrum, I am inclined to the opinion that this case is one of focal infection, the focus probably being in the posterior ethmoid cells, extending from the diseased antrum. The fact that the left eye was the first to be affected, whereas it was the right antrum that was diseased, is not uncommon. This contralateral visual disturbance in nasal sinus disease has been frequently observed by Onodi, as is stated in Knapp's *Medical Ophthalmology*. Whether the exploration of the ethmoid cells would have been of any benefit, as the process was so acute and so intense, is, of course, extremely doubtful.

This case illustrates to me a very wise remark our distinguished guest, Sir James McKenzie, made in New York, recently: "Let us not be exclusively Laboratory Physicians."

CATARACT IN RETINITIS PIGMENTOSA; ITS PATHOLOGY AND TREATMENT.[1]

By Dr. ARNOLD KNAPP, New York.

IT is a well-known clinical fact that a cataract frequently develops in the course of retinitis pigmentosa. Thus Leber[2] states that this cataract is so constant in the older cases that it often suggests a diagnosis of the condition. The cataract develops in the form of a star-shaped opacity at the posterior pole, limited to the posterior cortical layers. It remains limited to these layers for a long time and rarely develops into a total cataract. In the beginning there is a punctate opacity at the region of the posterior pole, to which one or more radiating opacities are joined, suggesting a star shape. These radiating opacities render the examination of the eye-ground difficult. This cataract is rarely accompanied by other forms of cataract formation; an associated opacity in the anterior layers has been described, and I have observed a well-defined rounded opacity just anterior to the nucleus in two cases of posterior polar cataract in retinitis pigmentosa.

This posterior polar cataract, though it advances but very slowly, seems to be greatly responsible for the reduced central vision of these patients. Doyne[3] was struck by the remarkable effect produced by the removal of the lens in retinitis pigmentosa, even when the posterior capsule is alone affected and not to a very great degree. Though it is generally believed that the prospect of sight after the removal of the lens is poor on account of the nature of the disease, Doyne states that this

[1] Read at meeting of American Ophthalmological Society, New London, July, 1918.

[2] Graefe-Saemisch, *Handbuch*, vol. vii., pt. ii.

[3] *Ophthalmoscope*, 1910.

is directly opposed to the facts. The improvement that occurs after the removal of the lens, even when so little affected that the normal observer can see every detail of the fundus without difficulty, this author regards as nothing short of marvelous. Doyne has performed the operation several times, and with one exception,—in which central vision was grossly affected,—he was struck by the improvement, which was out of all proportion to his expectations. His experience in this class of cases is so positive that he urges most strongly the removal of the lens in the presence of a posterior capsule that makes a difference in the examination of the eye.

After my attention was drawn by Dr. E. J. Curran, now of Kansas City, to Doyne's views on the advisability of extracting immature cataracts in retinitis pigmentosa, I have been on the lookout for these cases, and I now wish to report on four cases which have been operated upon by me.

CASE 1.—C. C., aged 65, was seen June 8, 1914, because the left eye had been failing for four years. Sight in the right eye was $\frac{20}{100}$; field was concentrically contracted to from 2° to 5° from the center. There was the usual posterior polar cataract in the left eye, together with a small opacity apparently on the anterior surface of the nucleus of that lens. The eye-ground presented the usual changes. Wassermann negative. The cataract was removed in the capsule without incident. After operation the vision with correcting glass was $\frac{20}{30}$. There were some opacities of the vitreous and an opacity on the hyaloid from a hemorrhage occurring at operation. Subsequent vision was probably better as soon as the opacity was absorbed, but it has been impossible to reëxamine the patient.

CASE 2.—M. S., aged 29, has never had good sight, especially for the last four years. Vision R.: $\frac{20}{100}$, L.: $\frac{20}{100}$. Field contracted to 5° about center, typical radiating posterior cortical opacity in the lens. The eye-ground presented the usual picture. Extraction with capsulotomy was performed, without accident, on the right eye; after a few days a severe iridocyclitis developed which resisted the usual methods of treatment and did not subside until three abscessed teeth in the upper jaw on the same side were extracted. The resulting thickened secondary cataract was operated upon about eight months later; vision at present with glass is $\frac{20}{30}$.

CASE 3.—I. S., aged 32, always suffered from night-blindness; the sight in the daytime has been failing for four

years. Vision is $\frac{8}{200}$ in each eye with the usual posterior cortical cataract and the fundus changes of retinitis pigmentosa. Combined extraction with capsulotomy, no reaction, followed by needling with a resulting vision of $\frac{15}{200}$. A thin after-cataract has re-formed, which, after another needling, should give still better vision.

CASE 4.—S. P., aged 52, has been troubled with night-blindness for many years. Gradual diminution of sight in the daytime. Vision equals $\frac{8}{200}$. Posterior polar cataract with advanced changes in the eye-grounds. Field very narrow. The right cataract was extracted in the capsule without event. The result of the operation was excellent, with perfectly clear media. With a correcting glass vision was only $\frac{8}{200}$. There was no optical impediment nor particular macular disturbance to explain this poor sight, except that the field was very small and the patient's mentality unusually dull.

To recapitulate, in these four cases—two of which were operated on by removing the lens in the capsule, and two with capsulotomy—the visual results were: 1, $\frac{20}{200}$ improved to $\frac{20}{70}$; 2, $\frac{20}{200}$ improved to $\frac{20}{60}$; 3, $\frac{8}{200}$ improved to $\frac{15}{200}$; 4, $\frac{8}{200}$ improved to $\frac{6}{200}$.

The method of operating required in these cases is not different from that of the usual cataract. The operation is not any more difficult. Doyne observed in every case that he had considerable difficulty in getting the lens away after opening the capsule, as if it seemed to be unusually adherent. I was not able to confirm this peculiarity in the delivery of the lens. After-healing is uneventful. There are no complications as far as the original eye condition is concerned. Doyne has observed that after extracting the lens the capsule has a tendency to form again; a film recurs in the pupil exactly like a wrinkled capsule that requires needling. The visual results are distinctly encouraging and I entirely agree with Doyne on the correctness of removing the lens under these conditions.

Pathology.—One of the cataracts extracted in the capsule was prepared for microscopic examination with the following findings. The capsular epithelium at the equator of the lens is wavy in outline, the cells are enlarged and have lost their outline; their protoplasm is granular. Underneath this layer there are irregular vacuoles and the adjoining nucleated lens fibers present vacuoles and are generally irregularly distributed.

At the epithelial boundary the new formation of lens fibers has taken place irregularly and to an increased degree. A thin band of elongated epithelial cells continues along the posterior capsule. Directly internal to this are nucleated cells which are vacuolated, distended, and have proliferated irregularly, often forming nucleated lens fibers. Just at the center and occasionally above and below the center there are irregular cavities filled with poorly staining lens detritus, albuminous material, globular bodies, and the edges of the bordering lens fibers are corroded; these cavities represent the lenticular opacities seen with the ophthalmoscope.

The changes therefore found in this form of cataract are changes in the equatorial capsular epithelium, prolongation of transformed capsular epithelium along the posterior surface, and irregular cavities of lens detritus in the posterior cortical layers.

The literature contains but few accounts of microscopic examinations of cataracts occurring in retinitis pigmentosa. The best description is that given by Wagenmann,[1] in which similar changes to those found in the above-described specimen are given.

[1] Wagenmann, "Pathologische Anatomie d. Retinitis pigmentosa," *Arch. f. Ophth.*, vol. xxxvii., 1891.

ANNULAR LENS OPACITY FOLLOWING CONTUSIO BULBI. THE REPORT OF A CASE AND A THEORY RELATIVE TO ITS FORMATION.

By Dr. PAUL D. BERRISFORD, St. Paul, Minn.

(With one illustration on Text-Plate XXI.)

VOSSIUS (1), in a session of the Medical Society of Giessen, 1903, was the first to call attention to an annular lens opacity occurring after ocular contusion. Three years later Keller (2) reported six cases and accurately described the lesion. Since that time, according to Casey Wood (3), some thirty or more cases of traumatic ring-shaped cataract have been recorded in the literature.

The following case not only presented clinically an annular lens opacity, but exhibited also a "macular hole," a lesion not so common as to render its clinical report *mal à propos*.

CASE REPORT

John S., an apprentice in an automobile shop, while assisting in the lighting of a welding apparatus, was struck on the left eye by a piece of hose, following an explosion.

The patient presented himself for examination January 4th, one hour after the accident. The left eye exhibited an excoriation of the skin of the lower lid, a small superficial laceration of the tarsus of the lower lid beginning at the free border of the latter, between its outer and middle third, and extending obliquely outwards for a distance of 7*mm* towards the retrotarsal fold, and a small laceration of the ocular conjunctiva between the limbus and the external canthus, with ecchymosis in and about the lesion; marked ciliary injection, cornea clear, aqueous turbid, vision of the right eye $\frac{20}{20}$, left eye $\frac{20}{120}$, pupils equal. Because of the

TO ILLUSTRATE DR. BERRISFORD'S ARTICLE ON "ANNULAR LENS OPACITY FOLLOWING CONTUSIO BULBI."

marked blepharospasm and general discomfort of the patient further examination was not attempted.

On January 6th, the patient complained only of poor vision in the left eye. Oblique focal illumination and the loupe disclosed no lenticular abnormality. By transmitted light could be seen a circular circumscribed lens opacity having a diameter of *3mm*, its center being situated slightly nasal to the corneal center, on or directly beneath the capsule. This annular opacity appeared to be made up of minute dust-like dots, some of which coalesced to form small masses. These masses seemed translucent, lardaceous in structure, and brownish in tone. This discrete ring appeared heavier and slightly broader in the superior temporal portion, least distinct in the nasally directed portion of the circle, being at no place broader than a hair. The area enclosed within the ring appeared clear excepting for a few scattered dust-like opacities situated chiefly in the superior temporal quadrant.

The fundus showed in exactly the macular region, a sharply circumscribed oval area occupying a transverse position, with a long diameter approximately one third that of the disk. This area contrasted strongly with the neighboring fundus, being sharply delimited, dusky red in color, and appeared to contain within it minute pigment deposits, as if the area were dusted with emery powder. At this time parallactic displacement disclosed no evidences of a surrounding elevated zone.

January 18th, the patient having resumed his duties, complained that he had difficulty in judging distances. Hering's test and bar reading substantiated his claims. Perimetric findings gave no accurate information regarding central scotoma. The vision of left eye $\frac{20}{20}$, with eccentric fixation $\frac{20}{20}$. The patient, refracted under atropin, gave the following results: O. D. + .50 S. + .50 C. Ax. 90 = $\frac{20}{15}$; O. S. + .75 S. + .50 C. Ax. 90 = $\frac{20}{20}$.

During the following weeks the patient was examined from time to time, the annular lens opacity appearing less distinct each visit, the nasal half disappearing first, later the temporal half, and finally the dots that were within the circle. The annular opacity completely vanished in seven weeks.

Twenty days after the injury the macular lesion presented the typical appearance of a "macular hole," a dusky red punched-out excavation surrounded by an œdematous halo. Later two grayish atrophic patches developed in the red depressed area which in the course of time increased in size. The "macular hole" was observed to change its contour and instead of remaining transversely oval, now appeared roundish by the extension of its superior boundary.

It was perhaps Knapp (4) in 1869 who first recorded a case of "macular hole." Since that time much attention has been given to the study of the macula following contusions of the globe, so that this condition now holds a well recognized place in ophthalmoscopy, its appearance being so striking that if once observed it will scarcely be overlooked.

ANNULAR LENS OPACITY.

Traumatic ocular injuries giving rise to an annular opacity of the lens may be accompanied by the following complications: Laceration of the lids, proptosis, extra-ocular palsies, subjunctival hemorrhages, conjunctivitis, superficial and deep opacities, erosions and perforations of the cornea, perforation and rupture of the sclera, prolapse of the vitreous, mydriasis, myosis, hyphæmia, gelatinous and fibrinous exudation into the anterior chamber, anterior and posterior synechiæ, ophthalmoplegia interna, rupture of the sphincter pupillæ, traumatic cataract, hemorrhage of the vitreous, commotio retinæ, retinal hemorrhages, optic neuritis, rupture of the chorioid, extensive pigment changes in the fundus as well as atrophic changes and "hole" in the macula.

The reported case was complicated by a laceration of the lower lid margin, laceration and ecchymosis of the ocular conjunctiva between the external limbus and the outer canthus, and a "macular hole."

The majority of cases thus far reported are the result of blunt force received from flying objects such as dirt, B. B. shot, footballs, broken drill punches, rocks, etc. However, Gifford (5), Casper (6), Natanson (7), Purtscher (8), (three cases), Holloway (9), Strader (10) have recorded instances due to perforating wounds all of which lay anterior to the lens with the exception of one case recorded by Purtscher (8). That this impact of the foreign body need not have its site on the anterior portion of the globe is illustrated by Steiner's (11) case. A typical Vossius's ring was observed after a shot in the temporal region, the bullet having passed into the orbit behind the eye, the latter remaining uninjured although displaced suddenly forward from behind.

Owing to the delicacy of outline of Vossius's ring, the len-

ticular surface requires a most careful inspection for its disclosure. There can be no doubt that many cases have been either overlooked, unrecognized, or not reported. The pupil should be dilated. Inspection by means of oblique illumination and the loupe is as a rule not sufficient. The author was unable to discover the opacity by this means. The binocular loupe and corneal microscope lend considerable assistance (Nicolai) (12). By means of the ophthalmoscope and the large concave mirror, at a distance of about 33*cm*, from the patient, the annular lens opacity could be plainly seen contrasting as it did with the red reflex of the fundus. Details were ascertained by using a small concave mirror and a + 20 lens.

The form of the annular opacity as the name implies is usually circular. Rarely is it oval in contour as illustrated in a case of Casper's (6) following a perforation of the cornea with a resulting prolapse of the iris, the ring being drawn out into a point downwards and laterally, corresponding to the defect of the iris. Höeg (13) has reported a case in which the circle was interrupted by clear lines. The lenticular opacity was bordered by a rounded corner corresponding to the upper former site of the temporal pupillary margin, consisting of an arc, the upper portion of which was slightly convex, almost horizontal, and a temporal portion, also slightly convex, but almost vertical. The breadth of the ring is variable. In some cases it is as thin as a hair, in others much broader. The annular opacity in an individual case not infrequently shows a lack of uniformity in both density and breadth, often appearing broader and heavier in its temporal half, the nasal portion appearing so faint as to be almost imperceptible. Such a clinical picture was to be observed in the reported case.

Close inspection in most cases shows the circular opacity to be made up of fine dots crowded, as a rule, closely together and in some instances accompanied by faint lines. These dots, it would appear, frequently coalesce to form small masses, the dotted appearance occurring as a rule in the faintest portion of the circular opacity. The circle is not always complete, occasionally the opacity being interrupted by clear spaces. In some cases the ring is pigmented as the reports of Vossius (1), Nicolai (12), Von Mertz (14), Löhlein (15), Robinson (16), Cates (17), and others will show. Pigmentation need not be

confined to the ring, but may appear outside (Robinson (16), Natanson (7)), or inside (Strader (10), Cates (17), Gifford (5)), the area enclosed by the circle. The case herein reported exhibited a few scattered dots situated in the upper temporal quadrant of the enclosure.

Löhlein (15) reported a case which on being viewed with a corneal microscope, showed a circle consisting of radially arranged dots and lines of various sizes extending inwards towards the center of the circle, giving the impression of an opaque disk with a distinct outline.

Hescheler (18) observed a cone-shaped opacity extending downwards from the circle extending to the periphery of the lens, and towards the upper nasal side another opacity the size of a pinhead.

Höeg (13) noted in one case a scarcely perceptible diffuse opacity within the ring, while towards the lower periphery of the lens there were several radial opacities which disappeared in a few days.

The transient character of Vossius's ring is a prominent feature, the opacity completely disappearing in from a few days to eight weeks. The reported case had a duration of seven weeks. The annular lens opacity does not impair vision to any marked degree although the complications of contusio bulbi which frequently accompany it often produce the opposite effect.

The mode of formation of annular lens opacity has been the subject for much discussion, Vossius (19) giving the first explanation: "By the indentation of the cornea following trauma, the iris is forced against the lens producing either an expression of pigment from the cells of the pupillary margin, which are fixed by fibrin on the anterior capsule of the lens, or as in the case of an unpigmented gray ring, the indented cornea forces the pupillary border of the iris against the anterior capsule of the lens producing a degeneration of the capsular epithelium at the point of contact." He bases his last assumption upon the researches of Schirmer (20) who produced experimentally a contusion cataract by the traumatization of the capsular epithelium, which after a time regenerated leaving no visible signs of injury. Through a small peripheral section of the cornea, Schirmer introduced a fine

blunt-pointed probe up to the opposite pupillary border withdrawing it in such a manner that the instrument passed gently onto the anterior surface of the lens. He regularly found a linear, occasionally a longitudinally oval opacity with vaguely defined borders, at the point of injury. When viewed with the loupe, it appeared to have a grayish color and situated beneath the anterior capsule. Occasionally the opacity was so faint that it was hardly perceptible, but when viewed by means of transmitted light was nearly transparent, and of a granular blackish appearance. The opacity appeared from 1½ to 12 or more hours following operation, its greatest intensity being observed after from thirty-six to forty-eight hours, when it began to decrease, disappearing in from a few days to a few weeks.

A clinical case which would appear to confirm the hypothesis that the cornea is brought in direct contact with the lens in the production of annular lens opacity, is one reported by Krusius (21). He found a faint gray opacity on the posterior corneal surface or on the corneal side of Descemet's membrane at a point corresponding to the opacity in the lens.

The theory advanced by Vossius, substantiated it would seem by the experimental evidence of Schirmer, might at first appear authentic. Nevertheless, clinical evidence renders the supposition untenable for many of the cases at least. To come straight to the point, the indentation of the cornea is not a requisite in the production of annular lens opacity. Small perforating injuries either anterior or posterior to the lens are known to have produced it as the cases of Gifford (5), Purtscher (8), etc., record. The site of traumatization need not necessarily be situated in the anterior segment of the globe as Strader's (10) case gives proof. A bullet lodged deep in the orbit pushing forward the bulb. Höeg (13) affirms that the displacement of the aqueous necessary to produce such a trauma that would bring the cornea and lens in contact over an area 3*mm* in diameter, could not possibly occur without rupture of the iris or cornea. It is his opinion that a small body applied with force to the center of the cornea would unquestionably result in a lenticular opacity more dense in the center than at the periphery. Certainly this is untrue, for often neither cornea center nor iris is ruptured, the bodies applying

the force varying from B. B. shot and steel chips to saddle horses, footballs, and missiles of clay.

Höeg (13) has advanced a second hypothesis as the causative factor in the production of annular lens capacity, which seems more plausible than that offered by Vossius. It is his belief that the ring is produced by a sudden rise of hydraulic pressure in the anterior chamber to such a degree that the iris is pressed forcibly against the lens. This theory would not be inconsistent with applied force from foreign bodies of various sizes and shapes striking at all angles.

Purtscher (8), Holloway (9), and Steiner (11) aver that in cases where pressure is applied posterior to the lens, the increased pressure in the vitreous drives the lens against the iris producing the annular opacity. Coats (22) refutes this assumption, stating that in his belief the production of the ring from pressure exerted behind the lens is quite impossible unless the iris becomes imprisoned between the lens and the cornea, for the iris will give away before the advancing lens.

Gifford (5) who at first accepted Vossius's explanation for the phenomenon later abandoned it. There came under his observation a patient who received a non-perforating wound of the inner limbus and adjoining conjunctiva of the right eye with a resulting annular opacity. "Here was a typical case of Vossius's ring resulting from a glancing blow of a small piece of copper at the inner periphery of the anterior chamber; an injury which could not possibly have pressed the center of the cornea against the iris." Gifford therefore adopted the more convincing hypothesis of Höeg.

Can we, through the theory of Höeg explain the presence of pigment granules within the ring as is not infrequently the case? This is the crux criticorum. The author believes this is the weak point in Höeg's assumption.

In the discussion of the causa causarum, operating in the production of this clinical entity the writer will confine himself to the lesion described as consisting of a ring-shaped lenticular opacity about 3*mm* in diameter composed of pigmented or non-pigmented dots the area within the confines of the ring or outside of it, in certain cases containing like dots. That transient opacities of the lens such as radial lines, streaks, stars, disk-shaped nebulæ may occur following contusions of

the globe is well known and has been observed in the presence of the above described lesion. It may be said with a fair degree of certainty that such opacities lie in the capsular epithelium or in the anterior cortex except in rare instances. On the contrary, pigmented opacities such as comprise Vossius's ring or areas adjacent to it can with consistency be said to lie upon the anterior capsule of the lens. If the above statements be true can we preclude the possibility that different pathological processes may be operating in each? The author believes we cannot.

Direct force applied to the lenticular surface after the method of Schirmer (20) or indirect violence brought about by beating the eye for several minutes (Roemer (23)) can produce experimentally these lenticular radii, nebulæ, etc. As far as the writer can ascertain, experimental evidence is wholly lacking to prove that such applied force mechanically results in an annular lens opacity and not the vascular phenomena that accompanies it.

It is the author's belief that a correct explanation of the pigment dots found within the area encircled by the ring is the problem which solves, in part at least, the question of formation of Vossius's opacity.

The plausibility of the assumption that by the increase of hydraulic pressure within the anterior chamber pigment is pressed from the iris and deposited upon the anterior capsule of the lens in the pupillary area must, in the opinion of the author, be received with doubt.

Gifford (5) holds that the disturbance of nutrition which the ring itself might produce may cause the dust-like opacities within the ring. Cates (17) logically contends that if the opacification of the tissue within the ring be due to a disturbance of nutrition produced by the ring, such an opacity would more likely appear of a uniform nature and not granular in character. He adds: "If we accept the premise that the ring, when presenting a brownish color contains pigment squeezed from the pigment epithelium of the iris, may we not, with reason attribute the occurrence of opacities of that color encircled by the ring to a simple migration of pigment particles from a like deposit in the ring itself ? "

The theories of Vossius and Höeg rest purely upon a mechani-

cal basis. In the formation of annular lens opacity may not another important factor enter into the discussion? In reference to non-perforating ocular injuries Parsons states: "The typical signs of cyclitis, more especially the presence of precipitates on the back of the cornea are seldom seen after contusions. Just as traumatic iritis in a greater or less degree follows such injuries, there can be no doubt that the ciliary body is also frequently involved. The haziness brought about by exudates in the aqueous must be attributed to iridocyclitis and there is little doubt that the vessels of the ciliary processes often suffer severely. It must be remembered that precipitates on the cornea are evidence usually of a subacute or chronic process, and this is generally absent."

Have we experimental evidence to warrant the above assertions? The pathological sequelæ of applied force to the eyeball is most beautifully exemplified by Bach's (24) work. He traumatized the eyeballs of rabbits with a lever apparatus producing a cloudy transudation in the anterior chamber with myosis ten minutes after the injury. The aqueous cleared and the pupil dilated after three quarters of an hour. Pathologically the anterior chamber was filled with a partly granular, partly filamentous contents, but only on the anterior surface of the iris and in the pupillary area, never at the region of the ciliary processes. Bach believes the cause of the transudation after traumatism is an alteration of the vessels of the iris, a paralysis, the consequence of contusion which led to extravasation of the albuminous substances of the blood. Microscopically he found the vessels of the iris very much distended and the iris tissue edematous.

It would, therefore, appear that after traumatic injuries of the eye of sufficient severity to produce an annular opacity, an iridocyclitis immediately follows, attested clinically by ciliary injection, myosis, cloudy aqueous, etc. Clinical and experimental evidence shows this in most cases to be of a transient nature, disappearing in a few hours at most.

That the dots comprising the ring and those within it in the observed case appeared in transmitted light, translucent, composed of pigment fibrin and not unlike the minutest K. P., formed in the mind of the writer a basis for a new theory. The deep ciliary injection and turbidity of the aqueous, both

of which were of short duration, led him to believe that the formation of annular lens opacity was of an inflammatory nature, a consequence of traumatic iridocyclitis.

The pathological picture presented by the anterior capsule of the lens may have had its origin in the following manner: As a consequence of the ocular contusion, a mild iridocyclitis results, an exudate from the vessels of the iris and ciliary body pouring out into the aqueous. As a consequence, the fibrin therein contained agglutinates the pupillary border of the iris to the anterior capsule of the lens. From the experimental evidence of Bach it is fair to assume that an attenuation of arrestment of the mobility of the iris of a temporary nature immediately follows the injury. When the iris regains its normal function, an action which takes place after a brief period of time, it frees itself from the fibrinous adhesion at its pupillary border, the fibrinous ring remaining at the point of contact.

How then are we to account for the pigmentation of the ring and the area enclosed by it? The coloration of the circular opacity may be derived from the pigment epithelium of the iris at its point of contact with the lens capsule or may arise from the following sources: uveal pigment contained within the leucocytes that have poured out into the aqueous or pigment derived from the destruction of red blood corpuscles.

The pigment within the area enclosed by the circle or beyond it may be the result of leucocytic migration from the ring itself as Cates (17) suggests, the deposition of pigment-containing leucocytes from the aqueous or pigment derived from the destruction of red blood corpuscles fixed by fibrin. The presence of macroscopical blood in the anterior chamber associated clinically with Vossius's ring is so frequently observed that the author believes this means should not be completely ignored as a possible etiological factor.

In conclusion the author wishes to state that for a more complete elucidation of this most interesting subject the following experimental studies should be attempted: 1. Striking various areas of the cornea and sclera by means of foreign bodies of divers sizes and shapes flying at different angles. 2. Notation of the intraocular pressure at the moment of contact. 3. The recording of microscopically obtained data showing clinically a typical Vossius's ring.

Until this is done we must content ourselves with the several theories advanced accepting tentatively the one which from a clinical aspect seems the most logical.

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CONCERNING THE ETIOLOGY OF HEREDITARY OPTIC NERVE ATROPHY. REPORT OF TWO CASES WITH INTERESTING X-RAY FINDINGS.[1]

By Dr. WM. ZENTMAYER, Philadelphia, Pa.

(With two figures in the text and six on Text-Plate XXII.)

IN this paper we are concerned only with the etiology of hereditary optic nerve atrophy. The cases here reported present nothing that is new in the symptomatology of the disease, and, as this phase of the subject is thoroughly digested in the monumental work of Wilbrand and Saenger, no essential purpose would be served by restating it.

The opportunity was recently afforded to make an X-ray study of the several members of two families in which the disease had made its appearance, and the results of this investigation are the feature of this communication. The findings are in no way conclusive but are sufficiently uniform to merit attention.

In the first family (C.) there were four children, three boys and one girl. One boy died of typhoid fever, the other two are affected. The girl is living and unaffected.

Case 1.—(J. C.), the first male, who was a stove molder, was affected when 30 years of age. He had the usual children's diseases but suffered no permanent sequelæ and has been in good health since. Smokes moderately and uses but little alcohol. In February, 1912, he accidentally discovered that the vision of the left eye was poor although he could see but large objects. The vision of the right eye was seemingly unaffected. By the latter part of August of the same year he had to give up work because of the poor vision in the left eye. Soon after this a blind spot appeared to the

[1] Read at meeting of the American Ophthalmological Society, New London, Conn., July, 1918.

outer side of fixation and then spread to the center. He was much annoyed by a continued shower of snow flakes which was finally replaced by a fog. Never had diplopia. When examined in November, 1917, eccentric V. R. E. = $\frac{1}{180}$; L. E. = $\frac{1}{2\,60}$. Iridic reflexes sluggish. There was an absolute central scotoma about 20° in extent. The optic nerves were greenish gray and the vessels contracted. The neurologic examination was made by Dr. W. G. Spiller and was negative except that the tendon jerks were over-prompt.

The nasal examination made by Dr. J. G. Schwerin showed; nasal cavities negative, tonsils and pharynx red and swollen. The X-ray examination made by Dr. Henry K. Pancoast showed an enlarged pituitary fossa both by deepening and in the anteroposterior direction, 13*mm* anteroposterior by 12*mm* deep.

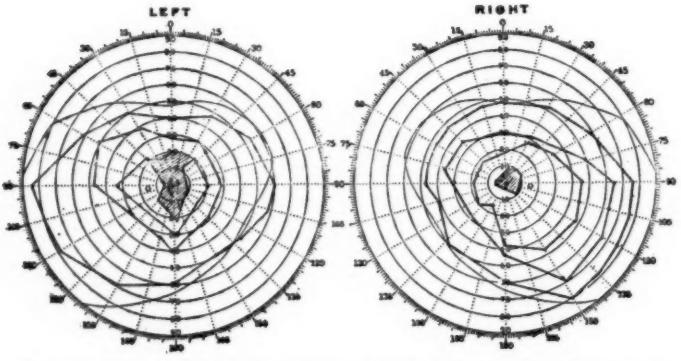


FIG. 1.—Case 2. Fields for form, blue and red. R. E., relative and absolute scotomata. L. E., relative scotomata.

CASE 2.— (C. C.), the second male affected, first seen in October, 1917, was 29 years of age. Until the summer of 1917 he had been employed in a glass works, the last seven years as a blower. During the summer, three weeks previous to coming to Wills Hospital for failing vision, he did outdoor iron painting. The paint contained Japan dryer. He had, however, no toxic symptoms. Three weeks after quitting this work he noticed that persons seen across a street looked blurred. Vision continued to fail until by the following week he could no longer read ordinary print. He thinks the sight of the left eye first began to fail, but of this he is not certain. He was in good health until the spring of 1917, when he had congestion of the lung with pleurisy.

He has smoked about 1 oz. of tobacco a day for several years. Does not use alcohol. He is married and has one child, living and well. His build is slight and he appears anæmic.

Pupils 4*mm*. Iridic reaction normal. Interpupillary distance 71*mm*. The upper nasal borders of the papilla are obscured and prominent. V. R. E. = $\frac{20}{100}$; L. E. = $\frac{15}{100}$. Visual field for R. E. shows a scotoma for red extending about 8° on either side of fixation and 20° below and breaking through the limit for red above. At fixation there is an absolute scotoma about 5° in extent.

In the field of the L. E. there is a relative central scotoma of about 5° extent. The form fields showed only slight contraction. It was thought that the condition might be due to an intranasal lesion and this was apparently confirmed by the examination made by Dr. J. G. Schwerin, who found both nasal cavities full of polypi and a purulent ethmoiditis. Complete eradication of these conditions, however, failed to arrest the progressive loss of vision but the œdema of the papilla disappeared. This was followed by an increasing atrophy.

A further search for the cause brought out the fact that a brother had lost his sight from optic atrophy (Case 1). The neurologic examination was negative. The aural examination by Dr. Lewis Fisher showed a hyperactive response in nystagmus and diminished responses to vertigo and past pointing, indicating a central disturbance. The X-ray examination showed possibly a slight deepening of the pituitary fossa—on the border line of normal size, anteroposterior 10*mm;* depth 11*mm*.

The second family (S.) in which this disease appeared consisted of nine children, five males and four females. Of these the fifth and seventh child, both males, are definitely affected, the third, a female, presents suggestive incipient symptoms of pituitary disease. The mother of this family was an only child. She is living at the age of 53 years. The father is 53 years of age and has normal eyes. There is no consanguinity in the ancestry so far as could be determined.

Case 3.—The first member affected (F. S.) is a male, now 26 years of age. At the age of 12 years the sight began to fail, and in six months' time he was no longer able to read. He thinks the left eye failed the faster. He was examined early in the attack by a competent ophthalmologist when a

diagnosis of optic atrophy was made. Vision has remained stationary in the last thirteen years. Except for scarlet fever at seven he has had no illness. At school he was a normal pupil until his sight failed.

Examined February, 1918; eccentric V. R. E. = $\frac{1}{120}$; L. E. = $\frac{1}{120}$, pupils equal, consensually right = 5.5*mm*, left, 5*mm*; R. E. slightly divergent. The face is asymmetrical, the right side recedes. The skull is high and narrow. The field of the right is contracted to within 40° of fixation, and there is a central absolute scotoma 20° in extent; the left is moderately contracted, and there is a central triangular absolute scotoma 60° x 15°. No color perception. Both

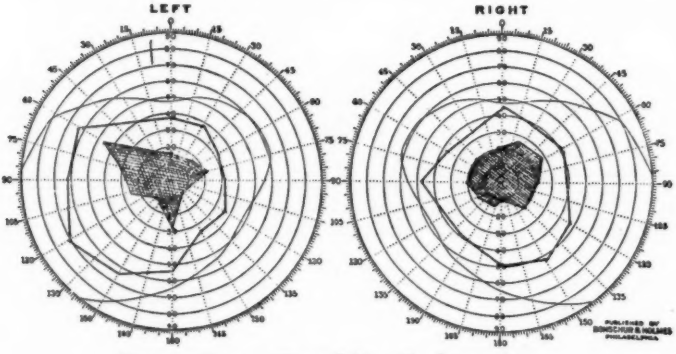


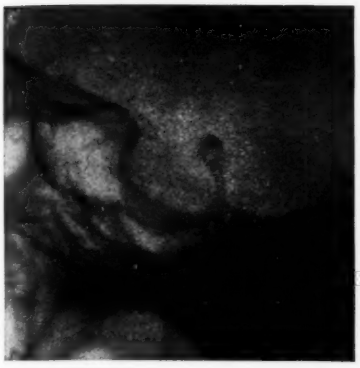
FIG. 2.—Case 3. Form field. Absolute scotoma.

optic papilla are atrophic with sharply defined laminæ. The vessels are of normal size in the right and slightly contracted in the left. The arteries are too bright. Under ophthalmoscopic examination the eyeballs develop a jerky movement usually up and to the right.

The physical and seriologic examinations are negative. The X-ray examination shows a large pituitary fossa on the border line of normal—12*mm* anteroposterior diameter and 10*mm* deep.

CASE 4.—The second male affected (E. S.) is 22 years of age, a chocolate maker for the past nine years. Two years ago he first noticed that colors did not appear normal, and examination of the eyes at that time is said to have shown an inflammation of the optic nerves. One year later he had repeated Wassermann tests, all of which were negative. These results were confirmed by our examination. The

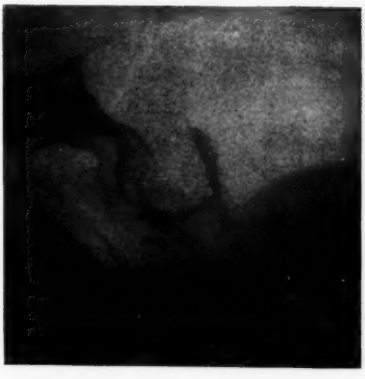
ILLUSTRATING DR. ZENTMAYER'S ARTICLE, "CONCERNING THE ETIOLOGY OF HEREDITARY OPTIC NERVE ATROPHY."



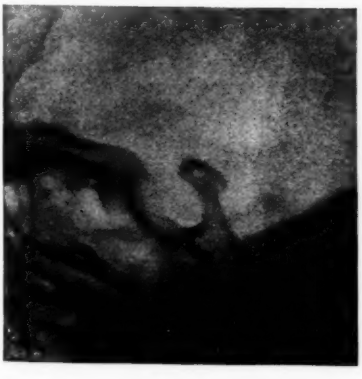
CASE 1.

CASE 2.

CASE 3.



CASE 4.



CASE 5.

CASE 6.

FIG. 3.—Cases 1, 2, 3, and 4 affected. Case 5, unaffected. Case 6, unaffected, but presenting indefinite pituitary symptoms.

nose, throat, and ears are normal. The Bárány test is normal. V. R. E. = $\frac{1}{60}$; L. E. = $\frac{2}{60}$.

R. E. The optic papilla, which is atrophic and is of a greenish white hue, has a deep pit-like excavation. The perivascular lymph sheaths are full. The vessels are contracted and the arteries too bright.

L. E. The papilla is gray and the vessels of a better size than in the R. E.

The X-ray measurements of the pituitary fossa are anteroposterior diameter 13*mm;* depth, 12*mm.*

The opinion of Dr. Pancoast that in cases 1, 2, and 4 the pituitary fossa was either above the average normal or at the maximum normal was confirmed when the cases were shown before the Philadelphia Roentgen Ray Society, and it was suggested that other members of the family who were apparently unaffected be examined as controls. Accordingly one male and one female of the second family were X-rayed. The male (C. S.) was 20 years of age and showed no ocular disturbance other than a high hyperopia. The pituitary fossa measured 9*mm* anteroposterior diameter and 9*mm* deep. The fossa was therefore much smaller than in the two affected members.

The apparently unaffected female (M. S. E.) is 30 years of age. The fossa measured 12*mm* anteroposterior and 10*mm* deep and is therefore on the borderline of being abnormally large. She subsequently came to my office for an ocular examination being fearful of a fate like that of her brothers. She is married and has two living girls aged 8 and 9 years. The last two children were boys, the first dead born, the second living but eight hours. She wore glasses at 14 years of age for a period of one year. She complains of momentary obscuration of vision, a great deal of headache, and annoying sweating of the left side of the head. Corrected V. = $\frac{5}{5}$ (normal).

The right papilla is normal, the left shows all but outer border obscured and slightly prominent. Central lymph sheath full. The visual fields showed decided concentric contraction for form and color with a minute central relative scotoma in the right field and an equally small paracentral relative scotoma in the left. Upon a second testing, five days later, on a Peter campimeter, no scotoma could be demon-

strated in the left field. She was examined neurologically by T. H. Weisenburg who found nothing abnormal aside from the left sided head sweating.

REPORT OF THE X-RAY EXAMINATION MADE BY DR. HENRY K. PANCOAST.

FAMILY C.: C. C.—At the first examination of this patient it was thought that the pituitary fossa was negative but after an examination of his brother, a re-examination convinced us that there was possibly a slight deepening of the pituitary fossa. This is about on the borderline of normal size.

J. C.—The pituitary fossa of this patient is slightly past the borderline of normal size and it is undoubtedly somewhat enlarged, both by deepening and in the anteroposterior direction.

REPORT OF THE X-RAY EXAMINATION OF FAMILY "S."

FAMILY S.: E. S.—The measurements of the pituitary fossa showed the fossa to be 13*mm* in the greatest anteroposterior diameter and 12*mm* deep.

F. S.—The pituitary fossa appeared to be on the borderline of normal. There is no evidence of any unusual pressure. The floor and posterior processes are thin but not out of proportion to the rest of the skull although thinner than in the case of the brother, C. S. The skulls of both C. S. and E. S. are of about the same thickness. The left anterior clinoid process is hollowed out beneath while the right is not. Both are normal but we usually do not find this combination. Measurements of the pituitary fossa—12*mm* anteroposterior diameter and 10*mm* deep.

C. S.—(One of the normal members of the family). The skull is thin. The pituitary fossa of normal size, and the processes about of normal thickness and apparently thicker than the two affected brothers. Measurements of the fossa, 9*mm* anteroposterior diameter and 9*mm* deep.

Mrs. M. S. E.—(Indefinite pituitary symptoms). The fossa in this case is large and on the borderline of being normal as in the case of F. S. The posterior clinoid processes and the floor are thinner than in the normal

brother. The skull is thicker than in any of the three brothers. Measurements of the fossa—12*mm* antero-posterior diameter and 10*mm* deep.

Dr. Pancoast concludes that as each of the four cases showed a pituitary fossa which was either at the borderline of being abnormal in size or was slightly past this point, it would seem as though a pituitary enlargement might be regarded as a possible cause of the condition.

Concerning the X-ray findings in the female who certainly has not, at present, an optic atrophy, but who has subjective symptoms, ocular fundus and field changes suggestive of chiasmal disturbance, it would seem not unfair to hold that at the present time it cannot be said to refute Dr. Pancoast's views.

It may be recalled that recently Fisher brought forward the following facts from which he argues an inherited temporary disorder of the pituitary body as the primary cause of Leber's disease.

Leber noticed a great tendency for the visual defects to appear at or about the age of puberty, and that evidence of a neuropathic type was afforded by such symptoms as headache, vertigo, tremors, numbness of all the limbs, or even epileptic fits; that in several reported cases there were similar symmetrical field defects; that patients with Leber's disease, as also those with rapidly developing pituitary body growths, often complain of subjective phenomena of light and colors often as "seen through a blue mist"; that variation in the degree of central amblyopia which occurred in Leber's disease is more consistent with an outside influence on the visual pathways than with primary changes in the nerve fibers; that in both conditions there is an epochal relation between the onset and the period of puberty and the climacteric and that in both, frequently in early stages of the disease, a very mild papillitis can be detected. In one of two affected children of a family, roentgenoscopy of the skull was negative, while the other showed a cellular or honeycombed shadow in the depression of the sella turcica. That similar changes were not found in the two cases he attributes to the fact that the negative finding was in a case of two years' standing. As the visual

symptoms in Leber's disease are progressive up to a certain point and then come to rest, he argues that if the lesion which gives rise to these symptoms is due to some disorder of the pituitary body it also must needs be temporary and transient.

Recently Pallock reports 2 cases of hereditary optic nerve atrophy each showing a small bean-shaped shadow within but below the center of the sella.

In regards to heredity, Harvey Cushing remarks that there may be certain inherited deviations which may in all likelihood be attributable to transmissible ductless gland properties; and a functional glandular instability may exist in these individuals which makes them more susceptible, under stress, to alterations which border on the pathological.

It would seem to me, that our data are insufficient upon which to draw conclusions. Aside from the fact that the normal fossa must vary greatly in size and often overstep the limits set as maximum normal, our present knowledge of heredity in connection with disorders of the pituitary body is too vague to permit of assertive statements.

I wish to express my thanks to Dr. Barry for his kindness in permitting me to publish the notes of the second family.

REPORT OF THE TRANSACTIONS OF THE SECTION ON OPHTHALMOLOGY OF THE ROYAL SOCIETY OF MEDICINE.

By Mr. HENRY DICKINSON, London.

An ordinary meeting of the Section took place on June 12th. Mr. William Lang, F.R.C.S., President, occupying the chair.

The President first referred to the decease of Mr. John Couper, a colleague with whom he had long been associated at Moorfields Hospital. He considered that Mr. Couper's chief claim to distinction was his special ophthalmoscope for estimating refraction by the direct method. It was this which made possible the late Marcus Gunn's minute studies, a work in which Mr. Gunn was worthily followed by Mr. Bardsley.

Dr. G. H. Goldsmith exhibited a case of **cystic swelling of the disk.** On the left side there was a large coloboma of iris and choroid. The disk was almost completely eclipsed by a swelling which he adjudged to be cystic. It was semi-translucent, and over the swelling could be seen two vessels.

Mr. F. A. C. Tyrrell showed a case of **Mooren's ulcer, in which the corneal denudation was extensive.** About a year ago, the patient attended St. Mary's Hospital with a large denuded area, the only epithelium being 4*mm* in the center of the cornea. As two attempts at cauterizing did not result in healing, he employed the conjunctival flap method, using purse-string sutures. The conjunctiva adhered to the denuded area in the outer part, and the cornea vascularized, but the case did not progress very well. At the end of a month, he determined to unite the lids in the center, in order to be able to flush the conjunctival cavity through from side to side. The cornea was now clearing up, and the patient could now

see fingers at the distance of a meter, and the pupil and iris could be seen.

The President said that in one case he touched the cornea with solid flavine, and though the patient complained of pain, it was very successful. Weaker applications were generally useful, but this was a more stubborn case.

Mr. LESLIE PATON, who had watched Mr. Tyrrell's case, confirmed its progressive character before treatment. It was the experience which had been gained in connection with sewing the lids in neuropathic keratitis which led to that method being adopted here. Clearing of the cornea followed equally well in cases of herpes in which the ulceration was deep. In the keratitis following operations on the Gasserian ganglion, however, the subsequent separation of the lids led, in his experience, to a relapse, unless at least a bridge of tissue of at least 1*mm* were left.

Mr. J. B. LAWFORD said his experience differed from Mr. Paton's in this respect, for a colleague of his at the hospital did a number of Gasserian ganglion operations, and Mr. Lawford sewed the lids together, and in not one of them was there a relapse when the lids were separated at a subsequent date.

Mr. A. ROXBURGH showed an **implantation cyst.** The patient, a girl æt. 14 years, had her eye struck by a tin toy aeroplane. The mother stated that the eye was operated upon immediately afterwards—probably at that procedure the prolapsed iris was removed. A fortnight ago the girl complained of pain, pricking, and headache. The mother then noticed a dark spot in front of the eye. Vision had fallen from $\frac{6}{9}$ to $\frac{6}{18}$, tension was slightly raised, but in the left cornea some iris was adherent to the back of the cornea. Beneath the cornea was a translucent cyst, about 4½*mm* wide, which appeared to be embedded in the iris, looking in every respect like an implantation cyst, and it was increasing in size. He felt it to be his duty to tackle it, but as he had not had such a case before, he asked for advice.

Mr. TREACHER COLLINS agreed with the diagnosis, and called attention to the importance of the cyst being removed, otherwise it would certainly fill the anterior chamber, and produce an increase of tension. The tissue removed at the

operation should include the portion of iris forming the back of the growth; if that were left, it would make practically certain subsequent extension.

The President agreed, remarking that these cysts often proved to be more extensive than was at first supposed.

Mr. M. S. MAYOU exhibited, on behalf of Captain Moxon, luminous test-types, arranged on the principle of the luminous wrist-watch, but of course with a special arrangement of lettering. An ordinary person in a completely darkened room could, after getting accustomed to the darkness, see these letters at a distance of four feet. Mr. Mayou had ascertained that the paint used for the purpose consisted of radium bromide and zinc sulphide, the light being produced by the bombardment of the zinc sulphide by the radium. It could not, however, very well be regarded as a standard, seeing that the degree of luminosity varied with the age of the paint, and with the thickness of the lettering employed. The scientific standard for estimating the light sense and the degree of night-blindness—which was the object of the device—would be a screen illuminated from behind, the observer wearing green glasses.

Mr. LESLIE PATON exhibited a patient in whom **chicken-pox was associated with neuroretinitis.** He had been able to find a record of only one other case of the kind, though neuroretinitis had occurred in association with poliencephalitis. There had been nothing else amiss with the present patient, and his blindness came on suddenly on the third day of his other disease. The large central scotoma left was now gradually clearing up.

Mr. C. P. BARDSLEY read a paper on a **new form of bi-focal for myopes.** He said there existed at the present day a school of ophthalmologists who taught that in early life myopia should be fully corrected, the subject wearing glasses to keep this up, however high the degree might be. This school argued that high correction only turned the child or young adult having myopia into a person with normal vision, and that thereby the accommodation was called into play, as was the accommodation of the emmetrope or the hypermetrope. The same people stated that if this full correction did not take place, certain dire results would ensue; accommodative power

would be lost, and there would be rapidly progressive myopia. But there had been no evidence adduced in favor of the idea that lack of full correction of myopia caused an increase of myopia. The author had himself been myopic since early childhood, and there had been only a slight degree of increase in it since that date, and when he was well over 40 years of age his accommodation was found, at the Ophthalmological Congress, to be more powerful than any so far recorded by that body. He asked why myopes should be robbed of the privilege of reading without using their ciliary muscles. On looking down through the lower glass in the ordinary bi-focals, there was considerable distortion of the image. To overcome this, the author angled his bi-focals; they were made of separate pieces of glass, slightly beveled against one another where the pieces met. Next, he angled the circular bi-focals, and to overcome the defect in bringing the edges of the upper and lower glass into continuous contact, he ground a toric curve on the upper lens, and a double concave or convex on the lower lens. He detailed the advantages resulting from this improved kind of spectacles.

Mr. L. R. YEALLAND read a paper on **hysterical disorders of vision,** based upon his experiences as resident medical officer at the National Hospital for the Paralyzed, Queen Square. He said hysterical disorders of vision could be grouped, clinically, into two classes. The first of these were conditions in which the contraction of antagonists could be demonstrated, and the second, those in which such was not susceptible of proof or demonstration. By contraction of an antagonistic group of muscles he meant the contraction of a group which should be relaxed during the performance of a given movement. This contraction of antagonists could be demonstrated in blepharospasm, ptosis, and spasm of accommodation. In blepharospasm there was simultaneous contraction of the levator palpebræ superioris and of the orbicularis oculi; failure of relaxation of the last-named muscle prevented the opening of the eye, hence there was apparent loss of vision. The action of the antagonists in spasm of accommodation could be explained on the assumption that the suspensory ligament of the lens performed an opposite action to that performed by the ciliary muscle. The suspensory ligament, held normally in a

state of tension, was only altered by the contraction of the ciliary muscle. When the latter contracted, the ligament relaxed; when the ligament was in a state of tension the ciliary muscle was relaxed. The phenomenon of contraction of antagonist could not be demonstrated in such conditions as limitation of the visual fields and amblyopia, but in these conditions contraction of antagonists in some other part of the body could be observed. The treatment was one of suggestion, and must be got through at one sitting. It produced complete relief of the physical disability, though much could not be hoped for in the way of permanent improvement in the mental state.

INDEX OF AUTHORS AND SUBJECTS.

VOL. XLVII.

Abducens Nerve, Traumatic Paralysis of the left, (H. H. Tyson) 204
Accidents, Wounds, Foreign Bodies, Parasites, Reports on, 112, 545
Accommodation, Spasm of, (J. W. White) 466
Achromatopsia, Total, (O. Wernicke), 302
Anatomy, Embryology, Malformations, Reports on, 427
Aniscoria: Two Cases of the So-called Physiological, or Loss of the Hereditary Correlation between the Two Halves of the Body, and Especially the Eyes, (Anton Lutz) 587
Aphakia at 48, (L. W. Crigler) 421
Argyll-Robertson Pupil, Report of Several Pathological Pupillary Cases, with Especial Reference to what Constitutes a True, (John Dunn) 469
Aviator, Eye Conditions Affecting the Efficiency of the, (W. H. Wilmer) 439

Bell, George Huston, Report of a Case of Bilateral Papillœdema, Due to Empyema of Sphenoid and Ethmoid Sinuses. Operation and Recovery, 344, 422
Berens, Conrad, The Need of an Ophthalmologic Clinic in Penal Institutions. With Special Reference to One Established at Sing Sing, 448
Berrisford, Paul D., Annular Lens Opacity Following Contusio Bulbi. Report of a Case of, and a Theory Relative to its Formation, 616
Blake, Eugene M., A Case of Evulsion of the Optic Nerve, 567
Blaskovics, L. v., The Operative Treatment of Traumatic Coloboma of the Eyelid, 404
Blind School, Results from 112 Wassermann Tests in the Missouri, (J. W. Charles and Harvey D. Lamb) 455
Boletin de la Sociedad de Oftalmologia de Buenos Aires, 295
Book Reviews, 211, 411, 436
Brain, Report of a Tumor of the Frontal Lobe of the, with Ocular Symptoms. Autopsy, (Dr. Arnold Knapp) 83
Brain and Hypophyseal Tumors, Distortions of Visual Fields in Cases of, and Chiasmal Lesions with Especial Reference to Homonymous Hemianopsia in, (Clifford B. Walker and Harvey Cushing) 119
Burge, W. E., The Production of Cataract, 12
Byers, Gordon M., Histological Findings in Eyeballs Lost through Complications Following Sclerocorneal Trephining, 462

Capsule, Organization of Lens Matter in the, (A. J. Herzig) 417
Carr, Andy M., Narrowing of the Pupil Does not Lower Normal Intraocular Tension, 177; Tension in Normal Eyes before and after Tonsillectomy, 46

Cataract, Microscopic Findings in a Case of Coralliform, with Remarks on Congenital Cataracts in General, (F. H. Verhoeff) 558; Production of, (W. E. Burge) 12; Suggestions for Improving Operations for, (Max Talmey) 425
Cavernous Sinus Thrombosis, Five Cases of, (Dorland Smith) 482
Chancre of Palpebral Conjunctiva, (A. S. Tenner) 422
CHARLES, J.W., and LAMB, HARVEY D., Results of 112 Wassermann Tests in Missouri School for Blind, 455
Chorioretinitis with Fixed Opacities in the Vitreous, (Uribe-Troncoso) 414
Choroid, Reports on Diseases of the, 534; Sarcoma of the, with Secondary Changes, (Edward Jackson and William C. Finnoff) 72
Ciliary Nerves, Herpes Zoster of the, (Thompson) 504
COHEN, MARTIN, Eye Manifestations in a Case of Polycythæmia, 192, 203; Leukæmic Manifestations in the Eye: A Clinical and Pathological Report, 61
Conjunctiva, Acute Streptothrix Infection of the, (Arnold Knapp and James G. Dwyer) 497; Contagious Diseases of the, Discussion on, 505: Reports on Diseases of the, 92, 520
Conjunctivitis, Pneumococcus, (R. Gil) 299
Cornea and Sclera, Reports on Diseases of the, 98, 526
Corneal Ulcer, (F. A. C. Tyrrell) 635
Cruise's Veil, 504

Disk, Cystic Swelling of Optic, (G. H. Goldsmith) 635
Divergence, Paralysis of, (J. M. Wheeler) 418
DUNN, JOHN, Headache as the Result of Eyestrain, 146; Report of Several Pathological Pupillary Cases, with Especial Reference to what Constitutes a True Argyll-Robertson Pupil, 469; Some Comments on the Probable Causes of Exophthalmic Goiter, 248; Some Phases of the Vestibular Nerve Problem, 354
DWYER, J. G., Focal Infection of the Eye from the Intestinal Tract, 261, 305

Ear and Eye, Intimate Relation between the, as Shown by the Bárány Tests, (H. M. Langdon and I. H. Jones) 348
Electric Ophthalmia with Chorioretinitis, (H. H. Tyson) 420
ELLIOT, R. H., "The Indian Operation of Couching for Cataract," Notice of, 211
Endarteritis, Obliterating, (White) 415
Epidiascope, (Gillies) 505
Exophthalmic Goiter, Surgical Treatment of Corneal Suppuration in, (Arnold Knapp) 173
Exophthalmos, Pulsating, (G. H. Bell) 417; (B. W. Key) 210
Eye, Extensive Perforating Wound of the, (B. W. Key) 204; Influence of Function upon the Structure of the, (E. Uhlenhuth) 401
Eye Diseases, Dental Infection in, (William F. C. Steinbugler) 182
Eyelids and Mastication, Associated Movements of, (Demaria and Caldora) 298
Eye Notes in War, (Collins) 501
Eyestrain, Headache as the Result of, (John Dunn) 146

FERGUS, FREELAND, Salvarsan and Other Remedies in Ophthalmic Practice, 29
Filaria Loa in Eye, (J. A. Cabaut) 295

General Ophthalmological Literature, Reports on, 310
General Pathology and Treatment, Reports on, 322
GIFFORD, S. R., Concerning the Value of the Blood Count in the Diagnosis of Sympathetic Ophthalmia, 327
GIFFORD, SANFORD, Leucosarcoma of the Iris Treated with Radium, 241
Glaucoma at 21, (L. W. Crigler) 421
Glaucoma, Iridectomy in, (J. H. Claiborne) 206
Glaucoma, Reports on, 105, 536
Goiter, Comments on Probable Causes of Exophthalmic, (John Dunn) 248
GREENWOOD, ALLEN; DE SCHWEINITZ, G. E.; PARKER, WALTER R., "Military Ophthalmic Surgery," Notice of, 437

HANSELL, HOWARD F., Ocular Affections Dependent upon Diseases of the Tonsils, Two Cases, 572
HARROWER, DAVID, Five Years' Experience with Iridotasis, 37
HINSHELWOOD, JAMES, "Congenital Word-Blindness," Notice of, 213
HOLDEN, WARD A., The Eye in Nervous Diseases, 221
HOLLOWAY, T. B., Asteroid Hyalitis, 50
Hyalitis, Asteroid, (T. B. Holloway) 50
Hydatid Cyst in Eyeball, (E. B. Demaria) 296
Hypotony in Rabbits' Eyes, (Collins) 502
Hysterical Disorders of Vision, (L. R. Yealland) 638

Implantation Cyst, (A. Roxburgh) 636
Intestinal Tract, Focal Infections of the Eye, from the, (J. G. Dwyer) 261, 305
Iridocyclitis, Experimental Study No. 5 in Ocular Anaphylaxis, (Alan C. Woods) 161
Iridotasis, Five Years' Experience with, (David Harrower) 37; Late Infection with Enucleation Following an Operation of, for Chronic Glaucoma, (Dunbar Roy) 42
Iris, Leucosarcoma of the, Treated with Radium, (Sanford Gifford) 241
Iris and Ciliary Body, Reports on the, 530
Iris and Pupils, Reports on the, 102

JACKSON, EDWARD, and FINNOFF, WILLIAM C., Sarcoma of the Choroid with Secondary Changes, 72

Keratitis Profunda, (J. Bruder) 201
Kerato Iritis and Teeth, (G. H. Bell) 210
KNAPP, ARNOLD, Cataract in Retinitis Pigmentosa; its Pathology and Treatment, 612; "Medical Ophthalmology," Notice of, 216; Report of a Case of Embolism of the Central Retinal Artery in Chorea, 459; Report of a Tumor of the Frontal Lobe of the Brain, with Ocular Symptoms, Autopsy, 83; The Return of Tension and the Occurrence of Retrochoroidal Hemorrhage after Trephining, 256; The Surgical Treatment of Corneal Suppuration in Exophthalmic Goiter, 173
KNAPP, ARNOLD, and DWYER, JAMES G., Acute Streptothrix Infection of the Conjunctiva, 497, 508

Lacrimal Passages, Reports on Diseases of the, 88, 518
LAGLEYZE, Notice of, 295
LAMBERT, WALTER EYRE, Acute Bilateral Retrobulbar Neuritis of Septic Origin, Resulting in Rapid and Complete Loss of Vision, 609
LANGDON, H. MAXWELL, and JONES, ISAAC H., The Intimate Relation between the Ear and the Eye as Shown by the Bárány Tests, 348
Lens, Annular Opacity of, Following Contusio Bulbi; Report of a Case of, and a Theory Relative to its Formation, (P. D. Berrisford) 616
Lens, Reports on the, 104, 533
Leukæmic Manifestations in the Eyes: A Clinical and Pathological Report, (M. Cohen) 61
Lid Lower, Gumma of the, (J. Bruder) 201
Lids, Reports on Diseases of the, 516
Lids and Mastication, 298
Lids, The Operative Treatment of Traumatic Coloboma of the, (L. v. Blaskovics) 404
Light and Color, Threshold of, (Young) 504
LUTZ, ANTON, The Light Pupillary Reflex, its Path, and its Abolition, Called Immobility of the Pupil to the Light Reflex, and Report of a Case of Unilateral Argyll-Robertson Pupil, in which Consensual Reaction Existed in both Eyes, 266, 370; Unilateral Inheritance. Loss of the Hereditary Correlation between the Two Halves of the Body, the Pair Organs, and Especially the Eyes. Description of Two Cases of the So-Called Physiological Anisocoria, 587

Macular Region, Congenital Coloboma of, (J. E. Weeks), 307

Miscellaneous Notes, Notices of Meetings, New Journals, etc., 218
MOORE, R. FOSTER, "The Retinitis of Arteriosclerosis, and its Relation to Renal Retinitis and to Cerebral Vascular Disease," Notice of, 411
Motor Apparatus, Reports on the, 433
Mucosa Transplantations, (R. Denig) 309
Myopes, Bi-Focal Lens for, (C. P. Bardsley) 637

Nervous Diseases, The Eye in, (Ward A. Holden) 221
Neuritis, Acute Bilateral Retrobulbar, of Septic Origin, Resulting in Rapid and Complete Loss of Vision, (W. E. Lambert) 609
Neuritis, Retrobulbar, with Tooth Infection, (W. D. Edwards) 513
Neuroretinitis with Chicken Pox, (L. Paton) 637
New York Academy of Medicine, Reports of the Proceedings of the Section of Ophthalmology of the, 201, 305, 414, 508
Nystagmus, Voluntary, (A. Gowland) 300

Ophthalmological Society of the United Kingdom, Report of the Annual Congress of the, 500
Ophthalmology, Reports on Progress of 88, 310, 427, 516
Ophthalmoplegia, Complete Bilateral, (A. Wiener) 307
Optic Nerve, A Case of Evulsion of the, (E. M. Blake) 567
Optic Nerve and Visual Tract, Reports on Diseases of the, 543
Optic Nerve Atrophy; Etiology of Hereditary, Two Cases of, with Interesting X-ray Findings, (W. Zentmayer) 627
Optic Neuritis, Cure of, of Accessory Sinus Origin, (S. T. Hubbard) 206
Optico-Ciliary Neurotomy, Resection of the Optic Nerve. A Substitute for Enucleation, (A. Pagenstecher) 409
Orbit, Report of Four Cases of Tumors of the, Successfully Removed with Preservation of Vision through an Orbital Incision along the External Orbital Rim, (W. C. Posey) 551
Orbit, Sarcoma of, (G. H. Bell) 209
Orbital Socket, Some Observations on the Restoration of the, (P. N. K. Schwenk and Wm. Campbell Posey) 576
Orbits, Exophthalmos, Accessory Sinuses, Reports on Diseases of the, 89, 519

PAGENSTECHER, A., Optico-Ciliary Neurotomy, Resection of the Optic Nerve. A Substitute for Enucleation, 409
Papillœdema, Bilateral, Due to Empyema of Sphenoid and Ethmoid Sinuses. Operation and Recovery, (G. H. Bell) 344, 422
Parinaud's Conjunctivitis with Tuberculosis, (P. B. Ferro) 299
Pemphigus Conjunctivæ, (E. M. Alger) 423
Penal Institutions, Need of Eye Clinic at, with Special Reference to Sing Sing, (C. Berens) 448
Plastic Iritis, (G. H. Bell) 209
Polycythæmia, Eye Manifestations in, (Martin Cohen) 192, 203
POSEY, WILLIAM CAMPBELL, Report of Four Cases of Orbital Tumors Successfully Removed with Preservation of Vision through an Orbital Incision along the External Orbital Rim, 551
Proptosis, Unilateral. Exploratory Operation. Tentative Diagnosis of an Ethmoidal Osteoma, (Robert Sattler) 68
Pupillary Reflex, The Light—its Path and its Abolition, with a Case of Unilateral Argyll Robertson Pupil with Consensual Reaction in both Eyes (Anton Lutz) 266, 509

Refraction and Accommodation, Reports on, 431
Relations of Ophthalmic to General Diseases and Poisons, Reports on, 315
Remedies and Instruments, Reports on, 323
Retina, Detachment of, (H. W. Wootton) 207; Report of Case of Embolism of Central Artery of, in Chorea, (Arnold Knapp) 459

Retina and Optic Nerve, Reports on Diseases of the, 109, 538
Retinitis Circinata, (W. F. C. Steinbugler) 513
Retinitis Pigmentosa, Cataract in; its Pathology and Treatment, (Arnold Knapp) 612
Retinitis Proliferans in Diabetes, (H. W. Wootton) 208
Retraction Movements of the Eyes, Congenital, (S. T. Hubbard and A. Duane) 416
RIVERS, W. C., "Consumption and Squint," Notice of, 215
Rosacea Ocularis, Accompanied with Fascicular Keratitis, (M. Uribe-Troncoso) 1
ROY DUNBAR, Late Infection with Enucleation following an Operation of Iridotasis for Chronic Glaucoma, 42
Royal Society of Medicine. Reports of the Ophthalmological Section of the, 195, 635

Salvarsan and Other Remedies in Ophthalmic Practice, (Freeland Fergus) 29
SATTLER, ROBERT, A Case of Unilateral Proptosis. Exploratory Operation. Tentative Diagnosis of an Ethmoidal Osteoma, 68; Penetrating Injury Limited to the Eyeball Followed by Acute Tetanus, 64
SCHWENK, P. N. K., and POSEY, WM. CAMPBELL, Some Observations on the Restoration of the Orbital Socket, 576
Scleritis, Probably Tuberculous, (F. N. Irwin) 208
Sense of Sight, Reports on the, 428
Seventh Nerve, Paralysis of the, (Santos Fernandez) 301
SHINOBU ISHIHARA, "Tests for Color-Blindness," Review of, 436
SMITH, DORLAND., Cavernous Sinus Thrombosis, with Notes of Five Cases, 482
Sphenoid, Tumor of, with Eye Symptoms, (Uribe-Troncoso) 510
Staphyloma, Ciliary, (B. W. Key) 205
STEINBUGLER, WILLIAM F. C., Dental Infection in Eye Diseases, 182
Sympathetic Ophthalmia, Reports on, 103, 534; Value of the Blood Count in the Diagnosis of, (S. R. Gifford) 327

Tension, Narrowing of the Pupil does not Lower Normal Intraocular, (Andy M. Carr) 177
Tetanus, Following a Penetrating Injury Limited to the Eyeball, (Robert Sattler) 64
THOMA, KURT H., "Oral Roentgenology," Review of, 436
Tonsillectomy, Tension in Normal Eyes before and after, (Andy M. Carr) 46
Tonsils, Two cases of Disease of the, Affecting the Sight, (H. F. Hansell) 572
Trephined Disk, Histology of the, (R. H. Elliot) 503
Trephining, Histological Findings in Eyeballs Lost through Complications Following Sclerocorneal, (G. M. Byers) 462; The Return of Tension and Retrochoroidal Hemorrhage after, (Arnold Knapp) 256, 308
Tumor, Orbital, Treated with Radium Successfully, (Bell and Tousey) 514

UHLENHUTH, EDUARD, The Influence of Function upon the Structure of the Eye, 401
URIBE-TRONCOSO, M., Ocular Changes in Acne Rosacea; One Case of Rosacea Ocularis Accompanied with Fascicular Keratitis, 1

VERHOEFF, F. H., Microscopic Findings in a Case of Coralliform Cataract, with Remarks on Congenital Cataracts in General, 558
Vernal Catarrh, (L. W. Crigler) 511
Vestibular Nerve Problem, Phases of the, (John Dunn) 354
Vision, Disturbances of, by Cerebral Lesions, Discussion on, 195
Vitiligo, Irido-Cyclo-Choroiditis in, (E. B. Demaria) 297

WALKER, CLIFFORD B., and CUSHING, HARVEY, Distortions of the Visual Fields in Cases of Brain Tumor. Chiasmal Lesions, with Especial Reference to Homonymous Hemianopsia with Hypophyseal Tumor, 119
WELLS, DAVID W., "The Stereoscope in Ophthalmology," Notice of, 437

WHITE, JAMES W., Spasm of Accommodation, 466
WILMER, WILLIAM H., Some Ocular Conditions Affecting the Efficiency of the Aviator, 439
WOODS, ALAN C., Ocular Anaphylaxis. V. Experimental Iridocyclitis, 161
Xanthelasma, Fulguration in, (Heller) 305
ZENTMAYER, WM., Concerning the Etiology of Hereditary Optic Nerve Atrophy. Report of Two Cases with Interesting X-Ray Findings, 627

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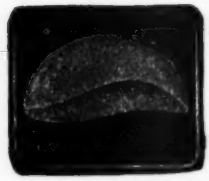

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